# Human Monstrosities

Barton Cooke Hirst

George A. Piersol

Part I.

# HUMAN

# MONSTROSITIES.

BY

BARTON COOKE HIRST, M.D.,

PROFESSOR OF OBSTETRICS IN THE UNIVERSITY OF PENNSYLVANIA,

AND

GEORGE A. PIERSOL, M.D.,

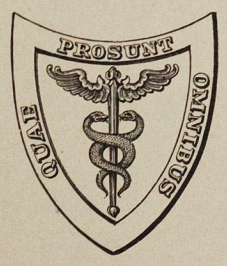
PROFESSOR OF HISTOLOGY AND EMBRYOLOGY IN THE UNIVERSITY OF PENNSYLVANIA.

## PART I.

ILLUSTRATED WITH SEVEN PHOTOGRAPHIC REPRODUCTIONS

AND

EIGHTEEN WOODCUTS.

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# PREFACE.

In the preparation of this work, the Authors have endeavored to present in concise and convenient form a treatise on Teratology which the practitioner can consult for an explanation of the graver anomalies in development encountered from time to time in medical practice.

The science of Teratology possesses more than a curious interest for the general practitioner and obstetrician who may at any moment see anomalies of development demanding recognition and treatment. To the anatomist and embryologist this study is of great importance, as the exceptions to the laws of development often emphasize and explain the rule.

The literature of the subject in English is widely scattered and often most unsatisfactory. We know of no English work comparable to those of Geoffroy Saint-Hilaire, of Förster, and of Ahlfeld, in which the entire subject is treated systematically, scientifically, and comprehensively. The physician who has been incited to a study of this science has been compelled, therefore, to seek information in foreign languages, and to devote much time to the perusal of works which, if comprehensive, are at the same time voluminous.

Fortunately, the materials are at hand for what we conceive to be a suitable volume. The Wistar and Horner Museum of the Medical Department of the University of Pennsylvania contains a rich collection of human monstrosities, some of the specimens in which are extremely rare and can be duplicated only in the largest European museums. Modern photographic processes have enabled us to transfer to the plates of this work accurate representations of many of the characteristic kinds of malformations. The numerous smaller illustrations throughout the text are gathered from many sources, some of which are by no means readily accessible.

If the work should in a measure fill a gap in English literature, and familiarize the public with the treasures of an American collection, the labor of the Authors will be requited.

Philadelphia, October, 1891.

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# PART I.

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# CLASSIFICATION.

Monstrous forms in the human species are the most radical departures from normal development presented by the offspring of man. They are the climax of anomalies in development. In attempting their description it is necessary to study systematically anomalies of development as a whole, for many monsters are but exaggerations of abnormal conditions frequently seen and of slight degree. The satisfactory classification of all anomalies in development is a task of no little difficulty; it must be remembered, however, that all classification is a matter of convenience, rather than an attempt to establish sharply separated divisions. The boundaries between some groups must be necessarily indistinct, as it is sometimes impossible to decide with certainty the heading under which a specimen falls.

We believe that the classification of Geoffroy Saint-Hilaire[1] is the most convenient for the physician who may meet examples of malformations and monsters in his practice, and may desire to locate and name them. Teratology is to the physician, in a measure, what botany is to the student of Nature. Both sciences should enable the investigator to recognize and designate strange and unusual growths. Geoffroy Saint-Hilaire's system is by no means satisfactory from the modern scientific standpoint. It takes no account of embryological laws or of

[1] Histoire générale et particulière des Anomalies de l'Organisation chez l'Homme et les Animaux, etc. Paris, 1832.

etiology, but in the present state of our knowledge it is impossible to construct an ideal classification, and we rest content, therefore, with one which is at least founded on wide observation, which groups together specimens similar in external characteristics, is flexible, permits of modification to keep pace with modern observation, and employs a nomenclature that has become familiar. The classification of Geoffroy Saint-Hilaire, therefore, we shall adopt—making, however, such alterations as seem desirable.[1]

The presence of any developmental abnormality in a human being places it in one of the following classes:

HEMITERATIC,
HETEROTAXIC,
HERMAPHRODITIC,
MONSTROUS.

[1] Many classifications of malformations have been proposed from time to time, those of Licetus, Buffon, Blumenbach, Meckel, Breschet, Gurlt, Otto, and Bischoff are given by Förster (Missbildungen des Menschen, Jena, 1861). See also Fisher, (Diploteratology, Albany, 1866); Ahlfeld (Die Missbildungen des Menschen, Leipzig, 1880).

## HEMITERATA.

I. ANOMALIES OF VOLUME.

*A.* Of Stature.

1. General diminution, as in a dwarf—delayed growth.
2. General increase, as in a giant—precocious development.

*B.* Of Volume, strictly speaking.

1. Local diminution, affecting—
   - *a.* Regions, as a limb.
   - *b.* Systems, as undeveloped muscles.
   - *c.* Organs, as small breasts, stenosis of canals, etc.
2. Local increase, affecting—
   - *a.* Regions, as the head.
   - *b.* Systems, as the adipose tissue.
   - *c.* Organs, as large breasts in women, lactiferous breasts in men.

II. ANOMALIES OF FORM. Single order, including—deformed heads; anomalies of shape in the stomach; deformed pelves, etc.

III. ANOMALIES OF COLOR.

*A.* Deficiency: complete, partial or imperfect albinism.

*B.* Excess: complete, partial or imperfect melanism.

*C.* Alteration: unusual color of iris.

IV. ANOMALIES OF STRUCTURE.

*A.* Deficiency in Consistency, as cartilaginous conditions of bones.

*B.* Excess in Consistency, as anomalous ossification, etc.

V. ANOMALIES OF DISPOSITION.

*A.* By Displacement.

1. Of the splanchnic organs, as anomalous direction of heart or stomach, hernias, exstrophy of the bladder, etc.
2. Of the non-splanchnic organs, as club-foot, curvature of spine, misplaced teeth, bloodvessels, etc.

*B.* By Change of Connection.

1. Anomalous articulations.
2. Anomalous implantations, as teeth out of line.
3. Anomalous attachments, as of muscles and ligaments.
4. Anomalous branches, as of arteries and nerves.
5. Anomalous openings, as of veins into left auricle, of the ductus choledochus in an unusual situation, of the vagina into the rectum, rectum into the male urethra, rectum at the umbilicus, cloaca.

*C.* In Continuity.

1. Anomalous imperforations, as of rectum, vulva, vagina, mouth, œsophagus, etc.
2. Anomalous union of organs, as of kidneys, testicles, digits, teeth, ribs, etc.; adhesion of tongue to palate.

*D.* By Closure, as in complete transverse septum in vagina.

*E.* By Disjunction.

1. Anomalous perforations, as persistence of foramen ovale, ductus arteriosus, urachus, etc.
2. Anomalous divisions, as splits, fissures in various organs, hare-lip, hypospadias, fissured tongue, cleft palate, fissured cheek, etc.

VI. ANOMALIES OF NUMBER AND EXISTENCE.

1. By numerical defect, as absence of muscles, vertebræ, ribs, digits, teeth, a lung, a kidney, of the womb, of the bladder, etc.
2. By numerical excess, as supernumerary digits, ribs, teeth, breasts, a double uterus.[1]

## HETEROTAXIS.

I. SPLANCHNIC INVERSION.

II. GENERAL INVERSION.

[1] As one regards this condition, it might be called an example of numerical augmentation or an anomaly by disjunction (failure of union of Müller's ducts). It is a good illustration of the difficulty of drawing hard-and-fast lines between the innumerable manifestations of anomalous development.

## HERMAPHRODITES.[1]

I. TRUE HERMAPHRODITES.

*a.* Bilateral hermaphrodites.

*b.* Unilateral hermaphrodites.

*c.* Lateral hermaphrodites.

II. PSEUDO-HERMAPHRODITES, with double sexual formation of the external genitals, but with unisexual development of the reproductive glands (ovaries and testicles).

*a.* Male pseudo-hermaphrodites (with testicles).

1. Internal pseudo-hermaphrodites. Development of uterus masculinus.
2. External pseudo-hermaphrodites. External genitals approach female type; feminine appearance and build.
3. Complete pseudo-hermaphrodites (internal and external). Uterus masculinus with tubes; separate efferent canals for bladder and uterus.

*b.* Female pseudo-hermaphrodites (with ovaries). Persistence of male sexual parts.

1. Internal hermaphrodites. Formation of vas deferens and tubes.
2. External hermaphrodites. Approach of external genitals to male type.
3. Complete hermaphrodites (internal and external). Masculine formation of the external genitals and of a part of the sexual tract.

[1] We substitute Klebs' classification of hermaphrodites as given by Ahlfeld, for that of Geoffroy Saint-Hilaire.

## MONSTERS.

### Class I. SINGLE MONSTERS.

#### Order I. Autositic Monsters.

- *Genus I.*
  - Species 1. Ectromelus
    - Phocomelus
    - Hemimelus
    - Micromelus
    - Ectromelus
  - Species 2. Symelus
    - Symelus
    - Uromelus
    - Sirenomelus
- *Genus II.* Single species, Celosoma
  - Aspalasoma
  - Agenosoma
  - Cyllosoma
  - Schistosoma
  - Pleurosoma
  - Celosoma
- *Genus III.*
  - Species 1. Exencephalus
    - Notencephalus
    - Proencephalus
    - Podencephalus
    - Hyperencephalus
    - Iniencephalus
    - Exencephalus
  - Species 2. Pseudencephalus
    - Nosencephalus
    - Thlipsencephalus
    - Pseudencephalus
  - Species 3. Anencephalus
    - Derencephalus
    - Anencephalus
- *Genus IV.*
  - Species 1. Cyclocephalus
    - Ethmocephalus
    - Cebocephalus
    - Rhinocephalus
    - Cyclocephalus
    - Stomocephalus
  - Species 2. Otocephalus
    - Sphenocephalus
    - Otocephalus
    - Edocephalus
    - Opocephalus
    - Triocephalus

ORDER II. OMPHALOSITIC MONSTERS.

| | | |
|---|---|---|
| *Genus I.* | Species 1. Paracephalus | Paracephalus |
| | | Omacephalus |
| | | Hemiacephalus |
| | Species 2. Acephalus | Acephalus |
| | | Peracephalus |
| | | Mylacephalus |
| | Species 3. Asomata | |
| *Genus II.*[1] | Single species, Anideus | |

CLASS II. COMPOSITE MONSTERS.

ORDER I. DOUBLE AUTOSITIC MONSTERS.[2]

A. Terata Katadidyma.

*Genus I.* Diprosopus.
*Genus II.* Dicephalus.
*Genus III.* Ischiopagus.
*Genus IV.* Pygopagus.

B. Terata Anadidyma.

*Genus I.* Dipygus.
*Genus II.* Syncephalus.
*Genus III.* Craniopagus.

C. Terata Anakatadidyma.

*Genus I.* Prosopothoracopagus.
*Genus II.* Omphalopagus.
*Genus III.* Rhachipagus.

ORDER II. DOUBLE PARASITIC MONSTERS.

| | | |
|---|---|---|
| *Genus I.* | Species 1. Heterotypus | Heteropagus |
| | | Heterodelphus |
| | | Heterodymus |
| | | Heterotypus |
| | | Heteromorphus |
| | Species 2. Heteralius | Epicomus |

[1] We omit the third order of Geoffroy Saint-Hilaire, single parasitic monsters, under which name he describes dermoid cysts.

[2] We substitute Förster's classification, with slight modification, for that of Geoffroy Saint-Hilaire.

CLASS II.—*Continued.*

| | | | |
|---|---|---|---|
| *Genus II.* | Species 1. Polygnathus | Epignathus | |
| | | Hypognathus | |
| | | Paragnathus | |
| | | Augnathus | |
| | Species 2. Polymelus | Pygomelus | |
| | | Gastromelus | |
| | | Notomelus | |
| | | Cephalomelus | |
| | | Melomelus | |
| *Genus III.* | Endocyma | Dermocyma | |
| | | Endocyma | |

ORDER III. TRIPLE MONSTERS.

# PRODUCTION OF MALFORMATIONS.

A CORRECT appreciation of the mode of formation of developmental anomalies depends so evidently upon an adequate acquaintance with the history of normal development that only with the advancement of our embryological knowledge has an intelligent study of malformations been possible.

The establishment of embryology upon a scientific basis of accurate observation by Caspar Friedrich Wolff (54), about the middle of the last century, laid the foundations for a rational teratology. The obscurity in which Wolff's second epoch-making communications lay buried unfortunately postponed the exertion of this important influence for more than forty years. When, at last, Wolff's researches secured the recognition which they merited, following their translation into German by Meckel in 1812, their powerful influence was exerted not only on current theories regarding normal, but likewise on those pertaining to abnormal, development; thus revolutionizing the accepted significance of anomalous formation.

What had been explained before by the most empirical speculation, or had been regarded as the expressions of Divine disfavor, or, perchance, as manifestations of the potent spell of some evil spirit, in the new light shed by the researches of Wolff, and of his apt pupils, Pander and Baer, now fell into the prosy category of deviations explicable by the application of the definite laws of development as set forth by embryology.

The important services rendered by embryology to teratology, in the infancy of both studies, have steadily accumulated and become more apparent with the extension of accurate, detailed knowledge of the various phases of normal development. Not only has embryology directly indicated the lines along which the production of anomalous development progresses, but it has emphasized the morphological relations of parts by pointing out the fact, that not a few deviations from the usual conditions in the higher types may be the regular and normal condition of lower forms.

While the increased knowledge so gained enables us to comprehend more satisfactorily the mode of formation of developmental deviations, and to better estimate their morphological significance, yet we must confess our ignorance of the primary causes producing many such variations, their etiology still remaining shrouded in uncertainty and speculation.

Those malformations arising from the mechanical effects induced by diseased and unusual conditions of the fœtus, as accumulation of fluid in undue quantity within the cavities of the bodies which directly influence the primary development, must be distinguished from the distortions sometimes observed as the results of inflammatory changes. These latter are to be regarded as the effects of pathological processes rather than as deviations of development.

Notwithstanding the paucity of our present knowledge concerning the predisposing causes, and of our ability to determine with certainty the etiology of many of the anomalies encountered, a general survey of their mode of production is not without profit, as aiding in the appreciation of the underlying principles involved.

Disregarding the foregoing classification, which from practical considerations has been adopted, malformations may be

grouped, with regard to their mode of production, conveniently into—

I. Those produced by variations in growth—either excessive or arrested.

II. Those produced by the defective union of component embryonic parts.

III. Those produced by cleavage (either partial or complete) of the primary embryonal cell-masses.

## I. Malformations Produced by Variations in Growth.

Excessive growth may involve all parts, producing a uniform increase of the entire organism, or be limited to a particular region, as one-half of the body, a single extremity, or even a single digit, as finger or toe. Variations from the normal length (50.5 cm. or about 20 inches) and weight (3250 grammes or about 7.2 lbs.) of the newborn infant are matters of every-day occurrence. Well-authenticated instances, however, of these figures being exceeded at birth to any remarkable extent are really of rare occurrence, notwithstanding the reputed frequency of such events on the authority of popular reports. A length over 200 cm. may be justly regarded as gigantic.

Individuals characterized by excessive growth quite frequently were of ordinary size during early childhood. The parents, likewise, are usually of ordinary size. A remarkable exception to this was reported by Beach (5) some time ago, where the father measured 7 ft. 7 in., the mother being 7 ft. 9 in. Their first child weighed 18 lbs. and was 24 inches long, while a second had a weight of $23\frac{3}{4}$ lbs., with a length of 30 inches, immediately after birth.

Regarding the causes leading to such unusual growth, absolutely nothing is known with certainty. Any attempt to refer

such excesses to the primary processes of development, by assuming the existence of an unusual amount of formative material, must be unsatisfactory, when we recall that in some cases the excessive growth began only after the ninth year, long after the cessation of embryonic activity. The exceptional nutritive activity producing these changes must, therefore, be recognized, without a knowledge of its cause.

Even more curious are those instances where only a limited portion of the body is affected by the excessive growth, as exemplified in cases where one extremity, a hand, or even a single finger, reaches inordinate dimensions, while the adjacent parts present nothing unusual. Care must be observed, of course, not to include within the category of "giant growth" parts enlarged by pathological processes, as inflammatory changes, elephantiasis, etc. While the affected part may be already, at birth, of unusual size, increasing by subsequent growth, it frequently happens that no such extraordinary character is to be noted, the local increase gradually developing after the expiration of some considerable time. The frequent participation of the osseous, vascular and lymphatic tissues in the hypertrophy indicates that the subcutaneous connective tissues alone are not the seat of the changes.

Anomalies due to defective growth may be considered under two groups:

*a.* Those in which a general decrease in size is accompanied by uninterrupted development.

*b.* Those in which diminution is dependent upon arrested or anomalous development.

To the first group belong the true dwarfs—beings characterized by completed physical development, although the resulting parts are of unnaturally small dimensions owing to insufficient growth. Authentic instances of very marked reduction in size

are of rare occurrence; sterility and impotence are not uncommon in pronounced dwarfs.

Regarding their etiology, nothing is known. Heredity seldom plays a part, since dwarfs are generally of parents of normal stature; they may at birth be distinguished by undersize, but more frequently attract attention only after the expiration of some time. True dwarfs are to be distinguished from individuals of diminished size consequent upon the effect of pathological conditions, as caries or inherited constitutional vices.

The second group, in which diminished size is due to arrested development and subsequent atrophy, includes very many various malformations, exhibiting all degrees of reduction, from a rudimentary finger to a shapeless acardiacus. The representatives of this group are produced by disturbed nutrition, the sequence either of general causes affecting the developing of the entire organism and resulting in general atrophy, or of localized impairment of nutrition, quite often mechanically produced as the effect of undue pressure exerted either from without or from within. It is highly probable that the general malnutrition of the embryo in the earliest stages, resulting in the production of those poorly developed or shapeless embryos occasionally encountered in the early abortions, is of much greater frequency than is supposed. His (23) has suggested the name "atrophic forms" for these malformations, which are characterized by want of general development, and, in some instances, by subsequent atrophy, whereby all resemblance to the human embryo is lost.

Conspicuous examples of arrested development due to impaired general nutrition are afforded by many specimens included within the group of acardiac monsters. The atrophic fœtus occurs associated with a well-developed, and frequently entirely normal, mate, the two usually possessing a common placenta in which two cords are inserted.

Opinions regarding the mode of production of these monsters are divided. The older view of Claudius (10) attributes the atrophic changes primarily to the reversal and impairment of the circulation consequent upon the overpowering vigor of the stronger fœtus. This doctrine has been warmly supported and further elaborated by Ahlfeld (2). According to this author, the formation of an acardiacus is largely a matter of accident. Ordinarily, the development of an ovum containing two embryonic areas results in the formation of homologous twins, characterized by great physical similarity.

The development proceeds regularly until the formation of the allantois—about the twelfth to thirteenth day; if the growth of the latter progresses with equal energy in both embryos, the available space for attachment offered by the primary chorion, or false amnion, will be equally divided, and each embryo will possess an allantoic, and later, placental circulation fully sufficient for its nutrition. The communication between the circulations established by the anastomosis of the allantoic capillaries, which is usually present in such homologous twins, exerts no serious influence on the development of either embryo, both completing normal gestation to be born fully capable of maintaining post-embryonic life.

Where, however, the anastomosis between the circulation of the embryos involves large vessels, inequality in the propulsive force may cause the gradual suppression of the normal blood-current within the vessels of the weaker fœtus, resulting in its death. The subsequent disappearance of the liquor amnii, obliteration of the amnionic cavity, and the pressing of the unfortunate fœtus against the uterine walls, follow as the effects of the increasing pressure consequent upon the growth of the amnion of the normal fœtus. The continued pressure flattens out the dead fœtus more and more, until, finally, the entire

mass has a thickness little greater than stout parchment; on account of these characteristic appearances, the name *fœtus papyraceus* has been given to such malformations.

In another group of twin embryos, a somewhat different fate befalls the weaker. When the allantois is formed with unequal rapidity in the two embryos, that of the more vigorous attaches itself, to the partial or entire exclusion of the less energetic fœtus. As a result, the latter is compelled to seek attachment for its allantois in the remaining unoccupied part of the area destined to form the placental attachment—the chorion frondosum.

Where, however, as sometimes happens, all of this placental area is already appropriated by the allantois of the first embryo, a secondary attachment to this united allantois is all that is left for the second fœtus to secure. Availing itself of this last source of nutrition, the weaker fœtus forms anastomoses with the allantoic or placental circulation of the stronger, thus becoming directly dependent for its nutrition upon the circulation of the latter, to constitute an allantoic parasite (Ahlfeld).

As a consequence of this dependence and of the greater blood-pressure in the vessels of the autosite, reversion of the blood-current in the weaker fœtus takes place, producing profound changes in many organs. The heart first suffers, losing its propelling function and becoming reduced in dignity to form but a section of the blood-conveying apparatus. This disappearance of the heart is the most constant characteristic of this class of malformations, although not necessarily so, since in certain cases the young heart may succeed in adapting itself to the reversed blood-current and become remodelled sufficiently to take part, partially at least, in the propulsion of the current; in such case a double circulation becomes established. Ahlfeld has called attention to this, and cited fourteen recorded acardiac monsters where a more or less rudimentary heart still existed.

In consequence of the usual absence of the heart, the anterior portions of the body are poorly supplied with nutrition, and, therefore, quite frequently cease to develop, undergoing atrophy resulting in monsters in which head, thorax, and upper extremities are wanting, or are present in an extremely rudimentary condition; such constitute the acephalic variety of acardiacus. Standing in marked contrast to this condition is the monster described by G. W. Koch (29), in which not only were throat, trunk, and the lower body surface apparently normal, but also the upper extremities were relatively well developed notwithstanding the entire absence of all, even rudimentary, traces of a heart.

The fresh blood from the umbilical vessels being directly poured into the aorta, those parts receiving their vascular supply from these vessels—as the lower extremities—suffer least, and quite commonly undergo a fair development. In those cases where the blood-supply is insufficient for the development of the lower extremities, the entire embryo becomes converted into a shapeless mass bearing little resemblance to the human form; such constitute the amorphous variety of the acardiac monsters. The hypertrophy of the subcutaneous connective tissue, which frequently follows the obstructed venous circulation in these malformations, still further contributes to the distortion of their external form.

Opposed to the foregoing views, championed by Ahlfeld (2), stand those of Dareste (12), Panum (34), Perls (35), Breus (9), Koch (29), and others, who hold that the reversed circulation and its sequelæ are but secondary, the primary changes being incidental to an inherent initial defect in the fœtus of so grave a character as to be incompatible with its further development. The life of such defective fœtus is continued by the timely aid obtained by the anastomosis of its allantoic circulation with that

of the more vigorous mate, thereby supplying the nutrition necessary to enable the fœtus to effect its partial development.

Regarding the respective claims of these opposed views, the truth seems to lie midway between them, in so far that the tardiness in acquiring allantoic attachments, or failure to maintain an independent normal circulation, depends upon a primary impairment of vitality of one, which renders the usurpation of the other fœtus possible.

The objection urged by Perls, that acardiac monsters have been encountered provided with special membranes and separate placenta, loses much of its weight when the possibility of aborted development of the heart from other sources is remembered: a fact emphasized by the observation of Dareste on the occurrence of an acardiacus in a chick without the presence of a second embryo. Whatever weight these criticisms may have in exceptional cases, there seem no sufficient grounds to reject, in its modified form, the doctrine of Claudius, as expressed above.

In the explanation of the production of those instances of arrested development and incomplete differentiation encountered in the extremities, in various degrees from the suppression of a digit to the fusion and rudimentary formation of the entire lower limbs (Siren), the question arises as to how far the arrest of development depends upon external, mechanical influences, in opposition to deficiencies of the formative material and forces.

Perls, Dareste, Gebhard (18), and others, agree in attributing such malformations to undue pressure induced by abnormal narrowing of the enveloping amnion. While limiting external relations no doubt induce important modifications, yet, when we recall the symmetry often seen in the defects in question, the fact that sometimes, as in phocomelus, the parts directly exposed to pressure are better developed than those more deeply situated,

as well as other features belonging to this class of malformations, it is difficult to escape from the conviction that the arrested development has its origin in a more deeply seated cause than merely mechanical opposition.

## II. Malformations Produced by the Defective Union of Component Embryonic Parts.

The second group of malformations, produced by the defective union or closure of the component parts, includes a long list of anomalies of very various degrees of gravity, from those causing but slight inconvenience, as simple hare-lip, to those incompatible with life, as extensive spina bifida.

These defects may be considered under two headings:

*a.* Those resulting from imperfect union of embryonal parts originally separated.

*b.* Those resulting from an imperfect closure of fœtal passages.

Recalling the primary embryonic processes, the fact is emphasized that the embryo is formed essentially by the development and subsequent union of two symmetrical halves. These halves, at first lateral tracts, by a process of proliferation and differentiation form two sets of folds—dorsal and ventral. The former unite above the line of the embryonic axis, as indicated by the notochord, to form a dorsal tube, the neural canal; the anterior or ventral folds join in front or below to form primarily the digestive tube, and, later, the anterior or ventral body-wall.

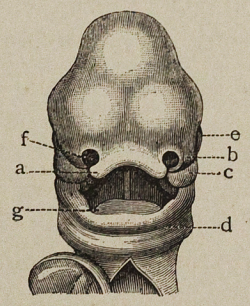
The dorsal line of union is unbroken, while the ventral line of closure is interrupted by the oral and umbilical primary openings, as well as by the secondary genito-urinary and anal orifices. From these facts it is to be anticipated, that anomalies

of defective union are of more frequent occurrence along the anterior than along the dorsal line of closure, a supposition corroborated by observation.

The immediate causes of the anomalies of this group, arising as the result of imperfect union, are very often mechanical, due to the obstruction offered to closure by the unusual presence of contiguous organs, on account of increased size, displacement, or abnormal attachments; sometimes, moreover, to the presence of an intruding fold of amnion between the uniting parts. In addition to the mechanical causes, formative inertia, associated often with insufficiency, or perhaps distortion, of the requisite material, is a potent factor in the production of ununited clefts.

The facial region, occupied as it is by the widely open primitive oral cavity, which obtains its definite form by the apposition and union of the five bounding processes, affords a

FIG. 1.




Head of a thirty-five-day human fœtus (after COSTE). *a*. Intermaxillary process. *b*. Lateral nasal processes. *c*. Superior maxillary processes of first visceral arch. *d*. Primary lower jaw, formed by the united inferior maxillary processes. *e*. Eye. *f*. Nasal pit and groove. *g*. Tongue.

favorite site for the occurrence of these anomalies. To facilitate the appreciation of the significance of developmental defects of the face, it will be advantageous to review the chief points in the normal method of closure. Reference to the accompanying

figure of a thirty-five-day human fœtus, after Coste, shows that the primary oral cavity is bounded by five processes—the broad central frontal or intermaxillary process (*a*), and the paired superior and inferior maxillary processes (*c*, *d*) of the first visceral arch. Between the outer nasal process and the superior maxillary process, lies the naso-orbital furrow leading from the nasal groove to the eye. The inferior processes earliest unite to form the inferior maxilla, this union being usually complete even when marked defects exist above. The superior border of the mouth is formed by the fusion of the inner nasal processes of the intermaxilla with the laterally placed superior maxillary processes.

When union of these parts fails to take place, total facial cleft results—a condition of which adhesion of the amnion to the embryo is regarded (Ahlfeld) as the direct cause. Such extended faulty union is infrequent, the defect being usually limited to one or more clefts. Imperfect closure along the naso-orbital furrow produces oblique facial cleft. Such anomalies may be unilateral; when so, they are more often seen on the left side. In explanation of this fact, the tendency of the embryo in the early stages to lie on the left side, has been assumed to facilitate the formation of disturbing amnionic attachments on the side in question. An unclosed naso-lachrymal furrow, forming an open lachrymal duct, also sometimes gives rise to an apparent oblique facial cleft. Where the intermaxilla fails to develop, while the superior maxillary processes almost meet, centrally cleft lip and palate result.

By far the most common defective union of the parts of the face is the lateral labial cleft, or hare-lip; while the defect may be limited to the lip, quite frequently it is associated with cleavage of the upper jaw and hard palate; much more rarely cleft bony parts exist without a corresponding fissure in the lip.

These deformities may occur only on one or on both sides; when unilateral, the left side is the most frequently affected. In addition to the mechanical influence of the amnion, heredity, as pointed out by Weber, must be accredited an important rôle in some, at least, of the defects of this group.

In enumerating the rarer defects of the oral cavity and the contiguous tissues, mention may be made of fissures of the cheeks, clefts, fistulæ and cysts of the lower lip, and defective inferior maxilla; all dependent upon obstruction to perfect union or to insufficiency of the formative material.

In the cervical region imperfect union of the body-walls along the ventral line gives rise to the central cervical fissure, associated with which tracheal fistula may exist. When, in the same region, the differentiation of the trachea from the œsophagus does not progress to complete separation, tracheo-œsophageal fistula results. Persistence of the visceral clefts gives origin to lateral cervical fistulæ. The question as to the existence of a thin epithelial partition separating the external furrow from the inner visceral pouch is still the subject of controversy. Many observers, advocating the opinion of His (24), deny the existence of a passage directly communicating with the pharynx from the exterior, unless artificially produced by rupture of the occluding membrane. Sutton (48, 49), however, records the observations of a case in which fluids swallowed appeared at the external orifice of the cervical fistule, although there had never been the possibility of an artificial puncture. Partial persistence of the inner visceral pouches gives rise to pockets in the lateral pharyngeal walls, which, in time, may become distended; cysts also arise from the dilatation of an isolated portion of the primary visceral clefts.

When the approximation and final union of the converging parietal plates is imperfectly accomplished, an external cleft

involving both thorax and abdomen results; defects of such extent, however, are infrequent, the fissure being usually confined to a limited area.

Ununited body-walls are met with in the thoracic region as sternal fissures, these being often the remains of larger clefts, which in the earlier stages involved the abdomen. Since abnormal volume of the heart must be regarded as a frequent cause of imperfect thoracic closure, cardiac ectopia frequently is associated with the sternal cleft, although the defect may exist independently as a rare anomaly. Defective development of the ribs, due to adherent thoracic parietes, sometimes causes lateral clefts through which a pulmonary hernia may protrude.

In connection with extensive thoracic cleft, imperfections in the diaphragm often exist. While possible at various points, such openings are most common in the muscular tissue of the left side. Portions of the small intestine, spleen, and other abdominal viscera may pass through these orifices. Reflections of pleura or peritoneum covering the migrated organs are but seldom present, the displaced viscera lying free within the thoracic cavity. The cause of diaphragmatic hernia, at least for those situated anteriorly, must be sought in the same conditions producing sternal cleft, rather than in an inequality between the pressure within the abdominal and thoracic cavities, since such sometimes assumed difference probably does not exist (Ahlfeld).

Imperfect union in the lower part of the thorax frequently is prolonged into the upper part of the abdomen; it is, however, the lower or posterior part of the ventral line of union, at the point of entrance of the umbilical vessels, that the greatest proneness to incomplete closure is manifested. Nor can this fact cause surprise, when we recall the various structures finding attachment to the embryo at or through this highway of communication between the embryo and the external sources of

its nutrition—at first by means of the vitelline circulation, and later through the placenta.

At first widely open, corresponding in extent to the walls of the large umbilical vesicle, with which the primitive gut is directly continuous, the abdominal orifice gradually narrows with the decreasing size of the umbilical vesicle and the formation of its narrow stalk. The amnion closely embraces both the umbilical and allantoic stalks, and, later, forms the external covering of the umbilical cord. At an early stage, when all its constituents are represented, a transverse section of the umbilical cord taken near the embryo discloses the presence of the vessels of the umbilical vesicle and of the allantois, together with the corresponding cavities of their stalks.

Such disposition favoring the incomplete union, the supervention of causes impeding closure, as amnionic adhesions, or undue bulk or displacement of abdominal organs, may readily lead to permanent fissures of the abdominal walls, with more or less extensive ectopia of the abdominal viscera. Imperfections of much less extent, however, involving only the umbilical ring, are frequent. Associated with the patulous condition of the ring, protrusion of the gut often exists, either as hernia of the cord, where a portion of the intestine, which normally lies within the umbilical cord until about the tenth week of fœtal life, fails to retract and remains without the abdomen; or, more rarely, as the result of secondary protrusion of the gut through the patulous ring, thus constituting post-fœtal umbilical hernia. All degrees of funicular hernia are seen, from the formation of the cylindrical diverticulum of Meckel, by the extension of the intestinal lumen at the point of attachment of the vitelline duct, to the displacement and inclusion within the hernia of large portions of the intestines and contiguous viscera.

Of the various explanations advanced regarding the produc-

tion of hernia of the umbilical cord, the one advocated by Ahlfeld is the most conclusive. According to this, the imperfect retraction of the gut depends generally upon abnormal attachments, due to persistence of the remains of the vitelline duct; in consequence of the continuous drag of the latter upon the intestines, the portion of the gut immediately attached is either actually drawn into the cord, or, as is more usual, it is merely prevented from retiring within the abdominal cavity as it normally should.

Depending upon abnormal position and consequent traction of the umbilical cord downward, a cleft involving the lower part of the anterior abdominal wall and pelvic bones sometimes is formed. Connected with this lesion, ectopia of the urinary bladder results from rupture of the allantoic sac. During the early weeks of embryonic life the contents of the gut, together with the excretion from the urinary glands, is emptied into the allantois, whose only opening is through the urachus. With the closure of this exit a new channel is established below into the cloaca; when, however, the closure of the urachus is not accompanied by the formation of a cloaca, the undue distention of the allantois from the accumulation of the effete matters poured into it, leads to its rupture in the direction of least resistance, which is toward the parietal walls, producing cleft bladder. The defect of the bladder is almost always associated with an abnormal adhesion of the gut, resulting in the formation of an artificial anus. A detailed and interesting discussion of the mode of origin of such intestino-vesico-abdominal malformations will be found in Ahlfeld's paper (3) as noted below.

As the result of the profound impression occasioned by the changed position of the parts affected in these malformations other secondary anomalies of development are induced. Among these are obstructed union of the Müllerian ducts, resulting in

double uteri and vaginæ, cleft conditions of the pubic region accompanied with faulty development and growth of the external genitalia. Following the displacement of the intestines an unusual space appears between the gut and the vertebral column; the occupation of this space is attempted by other parts of the intestines, as well as by the liver and kidneys. In certain cases the existence of this space induces inner and anterior curvature of the vertebral column, with secondary enlargement of the vertebral canal, and sometimes, ultimately, the production of spina bifida. The changes produced in the vertebral column may further lead to important secondary modifications in the formation and position of the lower extremities. Krüger (31) has described perineal hernia as a rare accompanyment of vesical ectopia.

It will be convenient to here consider the formation of those anomalies resulting from the imperfect union of the originally separated parts of the generative apparatus, the more so since, as already indicated, many of these variations are closely related to some of the incomplete unions already considered. Thus, the interposition of a gut attached to the bladder, as in cases of vesical fissure, or of a distended allantois between the two Mullerian ducts, prevents these tubes from fusing and results in the production of two separated generative passages, including oviduct, uterus, and vagina. All grades of such separation have been observed, varying from the completely isolated tubes, extending from the vaginal orifice to the oviduct, to the mere suggestion of division into double uteri by the splitting of the fundus. In addition to separation, asymmetry in the development of the two tubes not infrequently exists. This want of correspondence, even in normally united tubes, reaches its highest expression where one Mullerian duct remains rudimentary, while the other undergoes normal development, producing the unicor-

nuted form of uterus. Accompanying cleft symphysis, separated and imperfectly developed external genitals nearly always exist. Faulty union of those parts going to form the external genitalia, producing complete or partial fissure of the upper or lower surface of the penis or clitoris—respectively epi- and hypospadia—or divided scrotum, give rise to anomalies of interest on account of their conspicuous rôle in the production of supposed and so-called hermaphrodites. This group will be considered at length in later pages; however, it may be here stated that, notwithstanding the innumerable reputed hermaphrodites which have been described from time to time, there does not exist the unimpeachable record of a single case where the presence of true sexual glands of both sexes has been satisfactorily established. The case described by Heppner (25) is truly remarkable; the histological characters of the organs assumed to be testicles and ovaries, however, were not determined with convincing accuracy.

Malformations along the posterior line of union, due to imperfect fusion of the neural plates, are, as already stated, less frequent than along the corresponding ventral line. If the amount of intra-spinal fluid be increased, or the enclosing vertebral case be weak and defective, the contents of the vertebral canal will seek relief at those points offering least resistance. Such points will, naturally, correspond to those positions where union last takes place, where the knitted tissues are youngest and weakest, and where defective development is most conspicuous when present.

The parts of the medullary canal last to close are the lumbar and cervical regions, on account of the sharp flexures at these points, as explained already by His. Following as the sequence of this predisposition, the occurrence of defective union of the vertebræ and of spina bifida is more common in these particular regions than elsewhere, the lumbar, moreover, being

the favorite situation of such malformations. When the dimension of the extrusion which often appears is only moderate, the stretched and thinned-out integument forms an external covering to the sac; when, however, the tumor is of enormous size, as sometimes observed, the skin no longer suffices to envelop the sac, the sole covering being, under such circumstances, the distended membranes of the spinal cord.

Regarding the cause active in the production of spina bifida, opinions differ. According to the older and largely accepted view, increased intra-vertebral pressure, due to primary hydrops, is the most important factor, the ununited vertebral arches and the protruding sac being but the sequela of a force largely mechanical. An adherent amnion, preventing approximation and fusion of the neural plates, has likewise been regarded by some as the active element in the production of such defects.

v. Recklinghausen (40), on the contrary, after an elaborate study of the subject, concludes that these defects have a deeper-seated cause, originating in a primary aplasia of the skeletal axis. The interference with the normal development of the vertebral symmetrical halves hinders the fusion of the arches to form a simple tube. Such local malformations of the vertebral axis originate in an early defect of the blastoderm, as evinced by the associated imperfect development of other surrounding structures, including connective, muscular, and integumental tissues. When the developmental arrest affecting the spinal axis is expended upon those parts destined to become the bodies of the vertebræ, the rarer anterior form of spina bifida follows the anterior cleavage.

The symmetry of these vertebral defects shows that their origin must be referred to an early period of fœtal life, when deficiency of formative energy, as well as of material, was actively operative. Unequal and asymmetrical development of

the halves of the vertebræ, however, occurs in the production of mylocystocele, where the cleft is almost always laterally situated and to the left side. It is further of interest to note, that in addition to the asymmetry, mylocystocele is frequently, if not regularly, associated with intestino-vesico-abdominal cleft, both defects depending for their origin upon a common cause of arrested development.

The experimental researches of Richter (55), likewise, do not support the assumed importance of intra-vertebral dropsy or of amnionic pressure as primary causes of spina bifida.

Closely related to the malformations just considered are those affecting the cephalic expansions of the neural canal, giving rise to the group of monsters in which the vault of the cranium, together with the cerebral contents, is more or less defective. Regarding the mode of production of anencephalic monsters, opinions differ. According to the older view, supported by Dareste, Marchand, Perls, and others, the undue pressure and obstruction caused by an adherent and constricting amnion must be regarded as the most potent force in arresting development.

While admitting the possible influence of a constricting amnionic hood in the production of these defects, Ahlfeld holds the initial condition to be one of increased intracranial pressure, resulting in the rupture of the early cerebral vesicle, about the fourth week. The ragged edges of the opening resulting from this bursting gradually disappear, and, later, when these anomalies first are observed in the majority of cases, the cranial basis lies exposed, except where covered with overlying masses; these latter, on careful examination, prove to be composed largely of connective tissue, the remains of the investing cerebral membranes, together with knots of convoluted bloodvessels. Traces of the cerebral nervous tissue may also be present.

Opposed to these mechanical theories is the mode of the origin of the malformations under consideration, advocated by Ackermann (1), Quicken (37), and others, who attribute these deformities to defective development dependent upon insufficient primary growth or formative material—in short, upon some deeply seated central cause profoundly influencing development.

The close relation as well as the frequent association of these defects with those of the lower part of the vertebral canal, renders it more than plausible that the same forces are active in the impairment of development in both. The rudimentary condition often observed of the basal portions of the cranium and of the upper cervical vertebræ bears additional testimony to the influence of a condition of primary arrest of development.

In consequence of the imperfect formation of the base of the cranium the normal expansive forces are inactive, thereby inducing alterations in the relations of form and size of those parts forming the face, resulting in the physiognomy so characteristic of anencephalic monsters. When an ununited vertebral canal at the same time exists, a condition not infrequent, the occipital bone, if at all developed, is usually split. As a sequence of the same condition the cervical vertebræ are very rudimentary, so that the occipital region comes to lie in close proximity to the thoracic vertebræ. The face is, therefore, directed upward; this, together with the imperfect development of the orbits and the consequent apparent prominence of the eyes, produces that peculiar contour described as "frog-headed."

The anomalies depending upon the persistence of canals whose function normally ceases before or with the termination of intra-uterine life, while inconspicuous in their anatomical characters, nevertheless exert an influence often profound and sometimes incompatible with post-fœtal existence. The peculiarities of the fœtal circulation offer opportunities for persistent vascular

channels, among the most frequent being a patulous foramen ovale and pervious ductus arteriosus. The anomalies found in the large arterial branches near the heart are often referable to the persistence of early vessels usually obliterated; a double or transposed aortic arch finds explanation in the retention of parts of the aortic bows of the early visceral arches, which commonly atrophy. Retention to an unusual degree of the symmetry of the primary venous circulation explains such anomalies as a well-developed superior cava emptying into the coronary sinus on the left side, the duct of Cuvier with the primitive jugular having maintained their original relation instead of undergoing the customary partial atrophy. Persistent vitelline veins occur not infrequently—once in every hundred normal cords (Ahlfeld).

The fœtal communications between the gut and the extra-mural umbilical vesicle and the allantoic sac contribute frequent anomalies of greater or less gravity. The rôle played by persistent vitelline duct in the production of intestinal (Meckel's) diverticula, or, in more serious cases, of marked hernia of the umbilical cord and its secondary malformations, have already been considered. Persistence of the vitelline duct is dangerous on account of the possibility of the strangulation of the included intestine, as reported by Bradly and others. Where the lumen of the intestinal diverticulum extends into the cord, on separation of the latter an artificial anus may remain with all its attendant discomforts. Distention of a limited portion of the vitelline duct, which has become isolated by the closure of both the intestinal and the umbilical ends of the tube, may give rise to cysts.

With the formation of the bladder, and the discharge of the urino-intestinal secretions into the cloaca, the function of the allantois as a receptacle for excreta comes to an end; its duct, the urachus, becomes an impervious atrophic band, extending

from the fundus of the urinary bladder to the umbilical cord. Should, however, the natural channel of escape become obstructed, so that the contents of the bladder are not properly carried off, the canal of the urachus remains patulous, and the accumulations within the bladder are evacuated through the urachal orifice at the umbilicus. Sometimes, notwithstanding the closure of the urachus throughout the greater portion of its length, a part retains its lumen, and later may give rise to a cyst.

### III. Malformations Produced by Fission—either Partial or Complete—

Of the primary embryonal cell-mass;
Of the partially differentiated individual organs.

The malformations depending upon cleavage to a greater or less extent of the primitive early embryonic cell-mass constitute the most conspicuous group of anomalies, embracing all degrees of separation, from the striking instances presented by extreme double monsters, where union is maintained solely by a limited attachment, as between the sternum, pelvis, or head, to those slight fissions implicating a finger, toe, or internal viscus, producing supernumerary organs.

The mode of production of double monsters has long engaged the attention of eminent investigators; while opinions regarding the formation of multiple terata in certain points may be regarded as now settled, the views concerning many other questions are far from according. The older and once generally accepted theory, that double monsters arise by the fusion of two separated and independent products of conception, offered so many irreconcilable difficulties that it must be regarded as untenable and now obsolete, notwithstanding the once potent and

widely extended influence of the ingenious suggestion of the elder Geoffroy Saint-Hilaire, of the principle of the selection of like parts for like (*loi de l'affinité de soi pour soi*), a postulate upheld by the acceptance and teaching of his distinguished son.

While the view that all double monsters develop from a single ovum may be regarded as universally accepted, the details of this mode of origin are still subjects of doubt and controversy. Assuming that all multiple monsters arise from a single yolk—a view sustained by the most conclusive evidence—authorities are divided as to whether these monsters result from the union of two originally separate embryonic traces, or as the product of the more or less extensive cleavage of a single blastoderm. The former supposition recognizes primitive duality, followed by fusion (*Verwachsungstheorie*); the latter accepts primitive unity, later undergoing fission (*Spaltungstheorie*).

In addition to the authorities upholding the fusion theory in its older form, the production of double monsters by the union of two distinct primitive blastoderms on a single ovum has been maintained by B. Schultze, Panum, and especially Dareste, and has been provisionally accepted by Rauber and Gerlach.

Schultze (42) contended that double monsters sprang from ova possessing two germinal vesicles, the more or less perfect union of which gave rise to the more or less completely joined monsters.

Panum (34), likewise, concluded that the fusion of the two variously related primitive streaks is the cause of the malformations in question.

While the foregoing observers based their views on theoretical considerations, the investigations and conclusions of Dareste rest upon embryological investigations, and, therefore, merit especial attention. Dareste, while accepting the doctrine of

primitive duality, insists that the union can only take place in the early formative stages, and then only following a special preordination of the blastoderm. Fusion, in the sense of the older writers, of two embryos on separate ova, Dareste regards as impossible.

The occurrence of two blastoderms or cicatricula on a single yolk had been regarded as questionable, until the direct positive observations of Panum and Dareste, indirectly also Rauber, established the possibility of the existence of such double blastoderms on a single ovum beyond doubt. Gerlach also accepts the possibility of a bi-areolar development for birds, but regards the mono-areolar mode alone as applicable to the production of mammalian monsters.

While younger than its rival, the fission theory possesses a long list of illustrious supporters, including Wolff, J. F. Meckel, v. Baer, J. Müller, Valentine, Bischoff, and others—investigators whose names are inseparably associated with the foundation and advancement of modern embryology. The theory of fission, as presented by one of its most zealous supporters, Ahlfeld, accounts for all forms of double monsters. In consequence of unusual pressure from without, exerted by the enveloping structures upon the accumulated formative material of the embryo, the still undifferentiated embryonic cell-mass is cleft and displaced, this cleavage taking place prior to the appearance of the primitive streak. Where the pressure results in complete equal separation, the halves lie parallel and, at first, near each other, subsequently to be separated. Where the fission is incomplete only the divided portions change position on the blastoderm. Should the cephalic rudiments remain united, the caudal extremities diverge; should, on the contrary, the latter remain connected, then the anterior extremities become separated. By variations of the intensity of the action of these forces all

gradations between an almost parallel position of the two halves and the extreme divergences of head to head, or tail to tail, are possible. Ahlfeld (2) finds in fission the satisfactory explanation of all forms of double monsters, and regards as superfluous the aid of other theories.

Gerlach (19), on the contrary, considers the genesis of double monsters far more variable. This author, while accepting in the main the results of cleavage, does not admit the passive cleavage of the defined embryonic trace through the influence of external pressure, as assumed by Ahlfeld, but attributes to the embryonic trace an active participation in the process of division, the controlling impulse to which must be inherent within the embryonic cell-mass even before differentiation. This process of separation, for which Gerlach suggests the term "bifurcation," he regards as especially concerned in the production of anterior duplications; for the other forms of malformations, as posterior duplications, he recognizes the partial union or "copulation" of the two embryonic traces, which arise as separate but converging traces from the posterior margin of the germinal disc, after the manner of radiation.

The radiation theory, by which Rauber (38) seeks to furnish the explanation of the origin of double monsters, is based upon the fact that in the earliest stage the embryonic trace extends from the edge of the germinal wall or ridge toward the area pellucida as a radius. When a plural formation occurs Rauber assumes that two or even three of these embryonic traces appear instead of one, the development being designated as "pluri-radial." While Rauber's ingenious theory has the merit of resting upon embryological data, yet these are based so largely on conditions obtaining in the lowest vertebrates that we are not justified in attempting to directly apply it to the problems of the development of widely removed higher types.

In one other important respect Rauber's views differ from those of the advocates of the fission theory; according to the latter, a perfectly normal ovum, passing through a normal and typical fecundation and segmentation, gives rise to the embryonic mass which later becomes divided. Rauber, on the contrary, holds that a profound impression has been made upon the ovum, which determines from the beginning the exceptional development which the particular ovum will undergo; subsequent to segmentation, no force can affect the general arrangement of the embryonic cell-mass.

The former views have received support from the later investigations of Born (8), who, after the study of a large number of fish eggs, concludes that ova which produce double monsters pass through simple and regular segmentation.

Whatever the evidence in favor of the participation of other methods in special forms of monsters, it seems well established that division of the anterior end of the embryonal trace occurs as an expression of a far-reaching force in the production of anterior duplications. The lack of observations on early mammalian double monsters—the youngest human double monster being already four weeks old (Ahlfeld)—increases the difficulty of arriving at accurate conclusions regarding man and the higher types.

The early chick embryo of sixteen hours, which Gerlach (20) actually observed bifurcate, suggests the changes probably taking place in mammalian ova. The first thing noted was a broadening of the anterior end of the primitive streak; then, a forked divergence which became more pronounced, and by the twenty-sixth hour had reached a length of one-half of that of the undivided axis; from each anterior end of the diverging limbs a distinct head-process extended.

The acceptance as a fact, of the occurrence of the projec-

tion of two embryonic radii, with subsequent partial union, to form the posterior duplication, as advanced by Rauber and indorsed by Gerlach and others, offers some objections difficult to overcome. Notwithstanding the observations and arguments of Panum, Dareste, Rauber, Gerlach, and others, regarding the probability that in certain cases union and fusion of two at first separate cell-masses does take place in chicken eggs—especially since the occurrence of two blastoderms on a single yolk must be regarded as established—the objections pointed out by Ahlfeld (2) to the admission of the probability of the union of separated areas, still hold their force.

The well-established fact that, no matter how unequally nourished or how variable in extent, the union between the two halves of the double monsters is always accurately symmetrical, exactly the same parts being always joined together, seems difficult of explanation by the assumption that union of two separate areas has taken place, since such conditions would necessitate the uniform growth and unfailing approximation of the two areas, otherwise there would be observed occasionally want of symmetry of the union between the joined areas.

In the present state of our knowledge, then, we may assume that the bifurcation and divergence of the anterior extremity of the embryonic trace are largely accountable for the production of double monsters of man and the higher types; of the exact conditions and causes inducing this fission we know nothing. That, in certain cases, radial ingrowth and subsequent union of the separate areas may take place, is a proposition whose applicability to man and the higher vertebrates must be questioned.

Where the cleavage force is exerted to its fullest effect, the entire embryonal trace is completely divided; each resulting part may be capable of independent development, whereby two fœtuses are produced remarkable for their great similarity; such

offspring are known as *homogeneous* twins. They are always of the same sex and possess almost identity of physical characteristics.

In some instances, however, the cleavage force accomplishes but partial separation, resulting in the production of the conspicuous group of double monsters. As will be seen on glancing at the foregoing table of classification, all grades of bifurcation have been observed. The weakest expression of the cleavage force exerted upon the entire embryonic axis is manifested in the division of the anterior end of the notochord, giving rise to an anomaly which, if detected at all, owes its discovery usually to accident, since no outward peculiarity points to its existence. The most unrestrained action, on the other hand, short of the complete separation into two independent fœtuses, results in the xiphopagus, unless a bifurcated umbilical cord be regarded as the connecting form between twins and thoracopagus.

In the same manner that the embryonal area may undergo complete single cleavage, the resulting halves developing under favorable conditions into homogeneous twins, so also may a secondary fission of one of the halves take place, as an extremely rare occurrence, homogeneous triplets resulting from such two-fold division of the original area. Such fœtuses usually lie within a single chorion, enclosed by a separate or a common amnionic sac, the increased pressure arising from the unusual contents of the limited uterine cavity favoring absorption and disappearance of the amnionic partitions.

Instead, however, of the double cleavage resulting in the production of well-developed triplets, those causes already indicated as affecting twins, here still more often lead to the unequal development and the suppression of one or two of the fœtuses, which then become acardiac or amorphous parasites in connection with a single well-developed fœtus.

Just as in single cleavage, the double splitting of the primary area may be partial, involving, perhaps, but the anterior end, and producing thereby those rare tri-cephalic monsters, of which illustrations will be found in the succeeding pages. Again, the double cleavage may not progress in equal degree along with both planes of fission, resulting in the production of apparent double monsters, one of which shows secondary fission; thus there have been recorded monsters having two almost completely separated vertebral columns, one of which bore a single skull, while the second terminated in a divided cranium.

Multiple human monsters in excess of triplets are apocryphal; forms exceeding cleavage into triplets have never been observed. The development of two different varieties of fission in one individual must be regarded as one of the very rarest of all malformations. Ahlfeld (2) states that he found but a single case on record, that of Bongiovanni (7), where, in 1789, a living child was born showing a division of its face, and, at the same time, bore a well-developed dipygic parasite on the anterior thoracic wall.

While the extensively cleaved embryonic cell-masses may undergo uniform development, producing individuals capable of maintaining independent and almost separated existence—the celebrated Siamese twins being a noted example—not infrequently the halves of the bifurcated area develop unequally, resulting, sooner or later, in the predominance of the stronger part with a corresponding decline in the less fortunate member, whose circulation becomes more and more impaired, until from a perhaps former condition of equality the weaker part is reduced to dependency upon the stronger organism, to constitute a parasitic monster, of which almost all forms occur.

Since these double monsters usually come under observation in the later stages of their existence, the presence of an acardiac

or shapeless parasite, at first glance seems difficult of explanation by the theory of fission, until it is remembered that at one time both parts of the cleft embryonic mass were of equal dignity, and that subsequent unequal development is accountable for the discrepancy later observed.

When the development of two closely approximated embryonic traces is very unequal in the early stages, a series of curious and obscure parasitic monsters is produced by a process of inclusion by the parts of the more vigorous and rapidly growing fœtus, the imprisoned fœtus receiving its imperfect nutrition from its host. Where the two embryonic traces lie with their cephalic ends near together, in the course of the development of the more active embryo the weaker may be drawn within and overgrown or surrounded by the parts of the stronger, so that, finally, the only trace of the weaker fœtus is found as a tumor attached to some part of the head of the more vigorous fœtus, as the hard palate (epignathus), lower jaw (polygnathus), orbit, and other parts of the cranium.

A similar process may be instituted at the posterior end of the embryo, resulting in the formation of "congenital sacral" tumors, in the production of which a second included parasitic fœtus is the prime factor, although, at the time of examination the fœtal remains may have almost disappeared. An extension of this same process of surrounding growth by the parts of the stronger fœtus gives rise to those interesting inclusions to which the name "fœtus in fœtu" is sometimes applied.

Where the embryonic areas lie closely placed and parallel, the folding-off of the parietal plates—somatopleura—of one embryo to form eventually the anterior body walls, may be accompanied by displacement and dragging within the area space destined to become the body cavity by the future closing of the parietes; this displacement taking place, the rudimentary fœtus

undergoes its limited development more or less completely enclosed within the walls of the autosite, whose exterior in extreme cases may exhibit a slight tumor as the sole indication of the presence of an included parasite. On the other hand, the inclusion may be but partial, the parasite forming a conspicuous appendage on the body of the host; extreme examples of such rare anomalies are instanced by the cases of the Genoese Colloredo, the Chinese A-Ke, and others, which have become classic in the literature of teratology.

The included fœtus ordinarily lies within a sac, in the walls of which the remains of the enveloping amnion are usually to be found, together with a rudimentary umbilical cord. The sac is closely united with the adjacent organs whose bloodvessels contribute whatever imperfect nourishment the parasite receives. Should the displaced and included mass come to lie upon that part of the host corresponding to the germinal ridge destined to form the future sexual gland, it is possible for the imperfectly developed parts of the parasite to become surrounded by the tissues of the future testicle or ovary, thereby producing inclusio testiculi et ovarii.

These rare anomalies, however, must be distinguished from the relatively frequent dermoid cysts of these organs, in which various foreign tissues may be found. The difference to be noted is, that, in the true inclusion the parts are those of a partially developed entire organism disposed with some degree of regularity and order, and not, as in the case of the cysts, isolated, heterogeneous, usually ectodermic, tissues, representing but few organs. While the fœtal remains already found within the stroma of the testicle and ovary establish the occurrence of true inclusions within these organs as rare anomalies, yet the surmise of Ahlfeld, that the majority of such cases reported are to be regarded as really dermoids rather than true inclusions, appears well founded.

According to the same authority, the anomalies described as inclusions within the lung and mediastinum are to be regarded as dermoid cysts. In reference to the distinction between dermoid cysts and true inclusions of the cranium, Ahlfeld considers the difference in position as sufficient—when within the dura, to be regarded as inclusions; when without the dura, beneath, within, or outside the bony cranial wall, as dermoid cysts. Relating to the formation of these by no means infrequent anomalies, Ahlfeld emphasizes the possibility of some, at least, resulting from the "transplantation" of portions of a second fœtus.

The malformations formed by the limited cleavage of the cell-areas forming individual parts of organs, constitute an extended list, including almost every important organ of the body.

In explanation of the mode of production of deviations affecting the peripheral parts, as the limbs, the obstruction and opposition offered by abnormally adherent folds of the amnion have been assumed by some as a most potent cause. While, no doubt, the mechanical obstacles of adherent amnionic folds are capable of exerting, at times, important modifying influences, it must be recalled, that those parts most directly exposed at a time when the cell-mass is most easily impressed, as the early projecting limb-buds, show cleavage very rarely, if at all. Duplications of entire human limbs are doubtful; in the upper extremity splitting has never been observed, and in the lower limb only as the greatest rarity (Ahlfeld). Likewise, fission of the hands and feet is infrequent, while, on the contrary, division of the fingers and toes, and the production of supernumerary digits, are among the most common deviations.

Heredity, as well recognized, is conspicuously active as a cause of these anomalies, as the perpetuation of certain digital deformities through several generations shows; to assume in

such cases an inherited propensity on the part of the amnion to form obstructing folds can scarcely be regarded as a tenable proposition. Moreover, the occurrence of polydactylism among lower types, as amphibians, in which an amnion is never formed, shows that other forces than obstructing amnionic folds produce these anomalies. The symmetry often observed to a remarkable degree in these deformities also speaks for a more general cause.

It seems evident, therefore, that in some less localized and more deeply lying force must we seek the cause of these anomalies. As observed by Pott (36), the conditions of polydactylism, adactylism, and syndactylism are intimately related, being the variously modified expressions of the same force and process of differentiation, which, under normal conditions, produce the usual number of well-defined digits. Where this force is stimulated to unusual activity, by some influence exerted at an early period on the embryo, a supernumerary finger or toe results as the effect of changes affecting the cell-mass from within.

Excessive activity of the differentiating processes may result in the cleavage of the cell-areas destined to become other parts of the organism, including especially the ribs and vertebræ, muscles, vessels, and nerves, the more or less perfect duplications of which are often described as "variations" in anatomical text-books. In addition, very many organs occasionally present duplication, either as the result of primary fission of the early undifferentiated cell-mass, or, as perhaps is more frequently the case, of a secondary division of the partly differentiated organ, by the encroachment of neighboring structures, as bands of constricting mesodermic tissue.

Among the organs which occasionally present such cleavage, are the tongue, uvula, epiglottis, bronchiæ, lobes of the lung, stomach, intestines, liver, gall-bladder, pancreas, spleen, kidney,

ureter, urinary bladder, sexual glands, and mammary glands. Regarding the origin of supernumerary mammæ, two modes may be considered: Where the additional glands are found near the normally situated mammæ, a division of the primary gland area may be assumed to have taken place, with subsequent removal from the original site by the changes incident to growth; where, however, the supernumerary gland is encountered in localities far removed, as upon the thigh or back, fission of the original cell-mass offers an inadequate explanation, unless we assume that a portion of the original cell-area has been conveyed and transplanted, a supposition less plausible than to regard the appearance of such glands in unusual positions as the expression of a perverted differentiation of widely distributed epidermal structures, the mammæ being but the more highly differentiated specialization of the ectodermic tissue producing sebaceous glands. Supernumerary mammæ occur with almost equal frequency in the two sexes, the apparent greater frequency in women arising from the fact that the secretion of milk within the glands after childbirth quite often first calls attention to the existence of anomalies before unsuspected.

## THE ARTIFICIAL PRODUCTION OF MALFORMATIONS.

The interesting problems connected with the origin of double and other monsters, early suggested experimental investigations with a view of determining, if possible, the mode of production, as well as the causes and the conditions influencing their formation. While, for evident reasons, the attempts to artificially produce malformations have been limited to the ova of the lower types, the results are of interest as pointing out phenomena with a probable bearing on similar processes in the higher mammalian forms. In applying the deductions from such

experiments, however, the inherent differences between the totally segmenting alecithal mammalian ovum and the partially segmenting telocithal eggs of the fishes and birds, must be constantly borne in mind.

The possible causes influencing the production of monsters may be arranged in three groups:

I. Anomalous conditions of the sexual elements:
   *a.* Of the female element—the ovum.
   *b.* Of the male element—the spermatozoon.
   *c.* Of the union of the sexual elements, producing abnormal fecundation and segmentation.

II. Direct mechanical impressions.

III. Anomalous conditions of environment of the embryo, including thermic and respiratory changes.

For the purposes of artificially influencing development, it is evident that the first group of modifying conditions is beyond the reach of the experimenter. Born's experiments (8) with fish eggs showed that the ova of certain females were especially prone to yield double monsters, the tendency remaining almost constant and but little influenced by the difference in source of the spermatozoa. He also found that those ova which gave rise to double monsters passed through regular segmentation. That the ova of particular women may also possess an especial inherent tendency toward anomalous development is indicated by the repeated delivery of monsters from one mother.

On the other hand, it must be regarded as extremely doubtful whether peculiarities in the male elements are responsible for the production of monstrosities. The double-headed spermatic filaments sometimes seen, and to which the origin of double monsters has been ascribed, are variations occurring in

the development of the spermatic filaments themselves rather than cells possessing extraordinary fecundating powers.

While Fol (17), formerly also O. Hertwig (26), regards the entrance of several spermatozoa into the ovum as the signal for abnormal segmentation and development, the Hertwig brothers (27), after a more recent investigation, conclude that there is no proof that polyspermia produces double monsters. Likewise, the connection between double spermatozoa and the failure of the second polar body to be expelled with double monsters, as suggested by Windle (53), remains to be substantiated. So, also, the possibility pointed out by Dareste (16), that multiple pronuclei follow the penetration of more than a single spermatozoon.

The second group of modifying conditions—the influence of mechanical impressions—early attracted the attention of the experimenter. The mechanical influences employed by investigators in modifying the development of the ovum have been:

*a.* Abnormal position.

*b.* Disturbed equilibrium.

*c.* Direct mechanical injury.

The long-established fact that the germinal area corresponds with the lightest part of the egg, and, consequently, when unrestrained, always assumes a position on top, immediately beneath the shell, naturally suggested to the elder Geoffroy Saint-Hilaire (46) an investigation of the effects of a change of position. Although similar experiments had been undertaken by Reaumur, seventy-five years before, with negative results, the success of Saint-Hilaire in producing malformations by this method entitles him to the distinction of being the founder of experimental teratology.

Later, Liharzik (32) subjected eggs to change of position during the latter half of incubation, thereby obtaining chicks exhibiting distinctly the effects of the change in position. In

the normal ovum, during the second half of incubation, the chick lies with its axis parallel with the long diameter of the egg, the head being usually directed toward the broader end of the egg, the tail toward the smaller. Taking advantage of this relation, Liharzik placed eggs vertically, some with the broader end uppermost, others with it down. The effects of the augmented nutrition resulting from the increased blood-supply, aided by gravity, were distinctly apparent in the unusual size of that part which had been down, whether head or tail. Dareste (12) likewise obtained positive results by incubating eggs in the vertical position. More recently, Strahl and Gasser (45) succeeded in producing a partial inversion of the viscera by fixing the yolk with needles so that the germinal area lay beneath.

In another group of experiments, the disturbing influence was violent agitation. The early observations of Valentin (50) on the unusual frequency of double monsters in fish ova which had been subjected to violent shaking, were followed by those of Knoch (28), who obtained similar results by keeping the water constantly disturbed. The younger Geoffroy Saint-Hilaire (47) records that both he and his father retarded development by shaking eggs in a direction corresponding to their long axis; slight disturbances, on the other hand, as frequently turning the eggs during incubation, had no effect. Lombardini (33) produced numerous anomalies by causing the eggs to turn slowly about their long axis constantly for some days, at such a rate that a single revolution was completed in about one minute.

Dareste (13) found that eggs which had been greatly shaken by railroad transport, when incubated at once, usually die early; after several days of rest before incubation, however, eggs subjected to the same conditions developed normally. When shaken violently by a machine for some twenty minutes

before being placed in the oven, the eggs produced many anomalies, including cyclops, spina bifida, exencephalus, etc.; some eggs, in spite of all maltreatment, completed normal development.

Still more profound mechanical impressions in the course of development have been carried out by direct mutilation or injury to the ovum. Schrohe (41) split and punctured the germinal area with a view of producing double monsters; the results, however, were but deformed single embryos. So also Rauber's efforts to induce artificially the formation of divided embryos were negative; he therefore concludes, and with him Panum and Dareste, that it is impossible to produce double monsters by mechanical means. Similar experiments were carried out by Scymkiewizc (43), who incised the germinal area parallel with the primitive streak, and thereby obtained more or less distortion of the embryos. Puncture was also practised by Valentin and the younger Geoffroy Saint-Hilaire with the result of mutilations and arrested development.

In studying the effects of injuries to the amnion, Soboleff (44) removed a small fragment of shell and then tore or cut out a part of the amnion. Warynski and Fol (51) employed the thermo-cautery to make direct applications to the embryos. These authors conclude that cœlosomia and symelia are the only malformations for whose production the abnormal relations of the amnion can be held accountable. Gerlach (21) likewise applied the thermo-cautery to the anterior end of the embryo with the view of artificially producing exencephalus; his results were partially successful. Electricity in various forms as employed by Lombardini and Maggiorani (19) seems capricious in its action, although the disturbance and retardation of the normal development are, apparently, constant.

Experiments pertaining to the third group of influences, or

those affecting the supply of heat and of oxygen, have yielded most interesting results.

As long ago established by Reaumur, the most favorable temperature for development of the chicken's egg is about 40° C.; marked variations from this point, either above or below, when long continued are unfavorable for normal developmental processes.

Dareste, whose experiences in experimental teratology have been very variable and extended, found that at a temperature of 41° to 42° C., development progresses so rapidly that in twenty-four to thirty hours a stage is reached corresponding to that normally attained only at the third day; on the contrary, with a reduced temperature of 30° C., fully a week was required to reach a similar development.

By means of variations in temperature, Dareste (14) secured marked deviations in the form and development of the germinal, and especially the vascular, area. A continuous slight elevation of temperature retards growth while accelerating development, under which conditions dwarf chicks are frequently produced. Richter (55) obtained five malformations (three spina bifida with exencephalus and two exencephalus alone) out of seventy-eight eggs incubated at a low temperature.

Even more interesting have been the experiments by means of which the normal respiratory interchange of gases is affected. It is still undetermined, according to Gerlach, whether an excessive supply of oxygen induces alterations in development; on the other hand, that a diminution of the amount of oxygen reaching the egg seriously modifies its development is patent from the results of numerous experiments.

The effect of cutting off the access of oxygen by rendering the egg-shell impervious was studied long ago by Reaumur (39), who found that in eggs in which the porosity of the shell had

been destroyed, either by a complete coating of varnish or by being submerged in water, development did not take place. Further, that an atmosphere saturated with moisture was, for similar reasons, unfavorable.

Dareste's (11) elaboration of these experiments called attention to the fact that the coating must be applied immediately to the freshly laid egg, and be really complete and free from minute cracks to insure a total arrest of development when incubated. The disregard of these important precautions lead to apparently conflicting results, since, when the eggs lie for some time before coating, the supply of oxygen contributed by the air-space which is meanwhile formed, suffices for the developmental processes to a certain stage.

The experiments of Reaumur (39) in interrupting the development by a total covering of the shell, suggested to the elder Geoffroy Saint-Hilaire the possibility of producing malformations by limiting the supply of oxygen by an incomplete impervious coating of the shell. In eggs partly covered with wax this investigator obtained a number of malformations. Baudrimont and Martin-Saint-Ange (4) varied these experiments by coating eggs lengthwise, and found that when the coated area lay above, thus on the germinal area, development soon ceased.

Dareste employed various modifications of the coating methods in his experiments, and concluded that, as the result of the impervious covering of the egg the area vasculosa was less vigorously developed; the production of blood corpuscles being impaired, pronounced anæmia of the germinal area was induced. Under such unfavorable conditions of respiratory interchange asphyxia may readily occur. Dareste (15) also found that eggs incubated in air-tight spaces bear no direct relation to malformations. The age of the egg, however, seems to be a distinct factor in producing anomalies.

The investigations of Gerlach (19) in the domain of experimental teratology are of especial interest from the fact that his results, in yielding positive instead of negative data, differ from those of Dareste and others. Gerlach decided in favor of a partial impervious coating in making a choice of methods, with the special view of inducing, if possible, a cleavage of the anterior end of the embryonic trace and thereby producing double monsters.

Since the growth and proliferation of cells are always influenced by the supply of oxygen, Gerlach reasoned that by limiting the interchange of gases to a Y- or V-shaped area, corresponding to the anterior part of the germinal area, it might be possible to influence the disposition of the cells, by their search after oxygen, to such an extent as to cause an anterior bifurcation. Of sixty eggs so treated, nineteen exhibited variations which Gerlach believed stood in direct relation to the modified conditions. Among these anomalies were two unmistakable anterior duplications. Whether these two anomalies were really produced by the treatment to which the eggs had been subjected is by no means certain, since, as pointed out by Roux, such malformations occur in ordinarily incubated eggs much more frequently than Gerlach recognizes. One well-marked example of anterior duplication, of about fifty hours, was encountered in the last hundred eggs incubated by the author.

Gerlach and Koch (22), and afterward Koch (30), studied the effects of restricting the access of the atmosphere to the egg in the production of dwarfs, and found that a retarded development accompanied the diminished growth under such circumstances, resulting not in dwarfs, but simply poorly developed embryos. More recently, Gerlach (21) reports having artificially produced exencephalic embryos by limiting the supply of oxygen

by partially varnishing; in the same series of experiments the thermo-cautery was applied to the anterior end of the embryos, with partial success in producing exencephalus.

On reviewing the results of the foregoing attempts to produce malformations artificially, it becomes apparent that—direct mutilation disregarded—violent agitation, marked variations in temperature, and disturbance of the normal respiratory interchange, are all forces which, when acting on the early embryonic trace, are capable of producing profound alterations in the developmental processes not infrequently resulting in conspicuous malformations.

It is, however, equally clear that, as pointed out by Dareste, there is no definite correspondence or relation between the exact kind of disturbing influence and the character of the anomaly produced, other than the relation between a general disturbance of nutrition and the resulting retarded or arrested development. That by any given procedure we are able to produce at will a particular form of malformation is a consummation to which experimental teratology has not yet attained.

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# DESCRIPTION OF MALFORMATIONS.

## HEMITERATA.

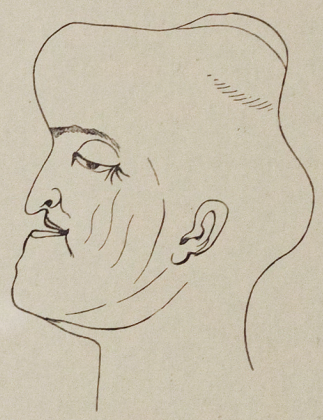
The hemiterata include all human bodies with anything unusual in development, not grave enough to be called monstrous nor of the specific character to be classed as heterotaxic or hermaphroditic. The difficulty of drawing a sharply dividing line between hemiterata and monsters in all cases is obvious. This is one of the weakest points in Geoffroy Saint-Hilaire's classification. For instance, an infant with a large umbilical hernia may be little removed from a celosomatic monster; and a cephalic meningocele may pass by easy gradations to a podencephalus. The difference is simply one of degree. But one can almost always decide, to his own satisfaction at least, whether the degree is marked enough to constitute a monstrosity. This, therefore, is a purely artificial division, not justified on scientific grounds, but we think it convenient to have a large general class into which all anomalies not of the specific character of the three other main divisions can be placed for further analysis.

It is manifestly impossible to refer separately to each anomaly of development that could be called hemiteratic in a work of this scope. Some of them, however, will be touched upon by reason of their clinical importance or scientific interest, before proceeding to a study of the monsters, to which a description of hemiterata, of heterotaxis, and of hermaphrodites is, in a sense, introductory.

Anomalies of Volume.—Many conditions come under this head. Some of them furnish indication for surgical interference, as macroglossia, unilateral hypertrophy of the face, etc.; or, on the other hand, narrowed canals of the body which require dilatation.

Anomalies of Form.—The deformities of the body most familiar and important to the obstetrician are found in the pelvis. There are many other examples to be found, however. A striking one is given in the illustration from Geoffroy Saint-Hilaire.

Fig. 2.



A deformed head (Egyptian).

Anomalies of Color.—Albinism and melanism come under this head. An individual with unusual color of the iris, or one in whom the eyes were of different color, would fall into this class.

Anomalies of Structure.—These are found mainly in cartilaginous or bony structures, but may be seen in the epidermis.

Displacement of the Splanchnic Organs.—This is an exceedingly large and important division, including as it does internal and external hernia, exstrophy of the bladder, etc. Some single examples under this head are so important that we shall glance at them in detail.

*Umbilical Hernia.* By this we mean, not the pouting of the skin over the umbilical ring, which is seen in about 2 per cent. of the newly born, but a true defect in the abdominal wall, leaving a space of varying extent around the umbilicus covered only by the bulging amnion, through which the abdominal organs can be seen, or through a rent in which they may protrude. In Plate I. is shown a very good example of umbilical hernia of moderate degree. Such a case demands the performance of a plastic operation, by which it could probably be cured. Of 19 cases so treated, recently collected, 17 recovered and 2 died. In 12 cases treated by non-interference, there were 9 deaths and 3 recoveries.[1]

*Meningocele.* A tumor composed of cerebral or spinal membranes containing fluid, and perhaps nerve tissue, protruding from an anomalous opening in the cranium or spinal column,

FIG. 3.

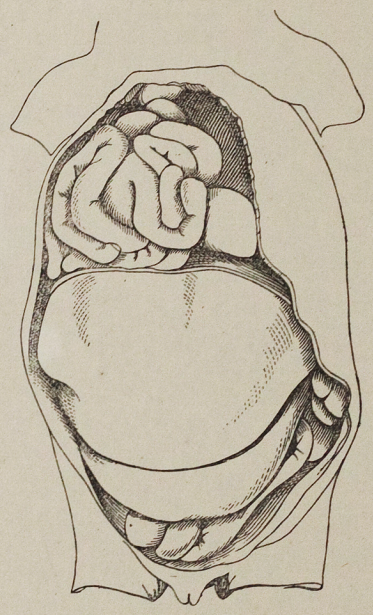
Meningocele at the root of the nose.

usually covered by skin, but sometimes exposed by cutaneous defect. In the cerebral variety one may often be puzzled where to place the case, whether among the hemiterata or monsters. This is well illustrated in Plate III., and to a lesser degree in

[1] MacDonald: American Journal of Obstetrics, Jan. 1890.

Plate IV. There should be no difficulty in making the diagnosis of such a condition, but we have known a cephalhæmatoma to be called a meningocele, and a meningocele to be called a hernia of the longitudinal sinus. The spinal meningocele associated with spina bifida may reach an enormous extent. A very large tumor of this sort, laid open, is seen in Plate II. Various plans of treatment have been proposed for this condition. The sac has been punctured; irritating substances have

Fig. 4.



Diaphragmatic hernia.

been injected; the whole sac has been dissected out, ligated, and cut off. A plastic operation on the bones of the spine themselves has been suggested in order that the opening into the spinal canal might be permanently closed. But the liability to inflammation of the spinal meninges, to fatal brain disorder from intra-cranial pressure, to septic infection, and the likelihood that

hydrocephalus exists, make the success of any plan of treatment doubtful. We believe, however, that the dissection of the sac, avoiding nervous tissue, and the ligation of its pedicle under strict antiseptic precautions, will most often succeed.

*Diaphragmatic Hernia.* This hernia is characterized by an abnormal opening in the diaphragm, through which the abdominal or thoracic organs pass into an unnatural situation. The diaphragmatic opening is usually upon the left side. The hernia may not occur until after birth, when the action of the abdominal muscles may so increase the intra-abdominal pressure as to drive the abdominal contents upward through a preëxisting fissure in the diaphragm. This fissure is more often found in the muscular than in the fibrous portion.

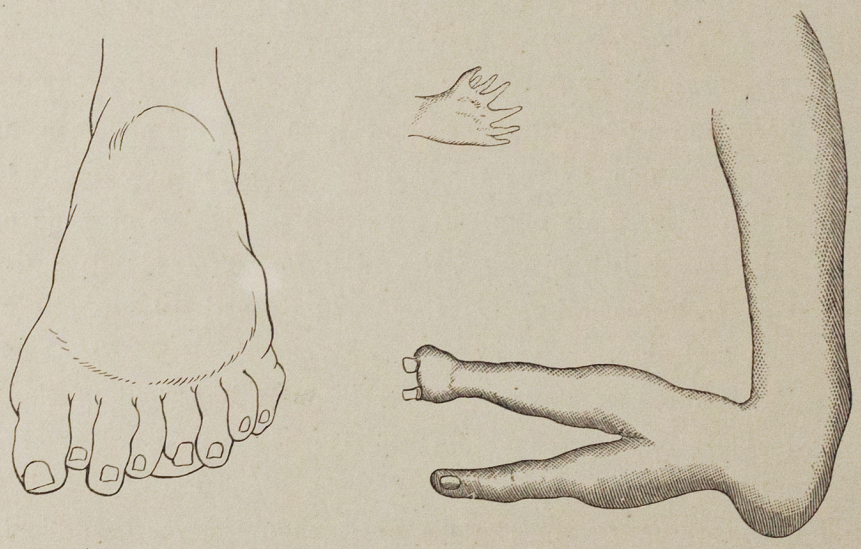
*Displacement of the Heart and Lungs.* The heart may be displaced upward into the neck, downward into the abdomen, in various directions in the thoracic cavity, or it may protrude through a fissured sternum. As a result of absence of some of the ribs, especially the upper, the lung may protrude from the chest at each respiratory effort, covered by nothing but pleura, muscles, and skin.

Anomalies in Continuity.—The most important of these are, perhaps, imperforate rectum and œsophagus. An imperforate rectum may be remedied by operations, which are described in all works on surgery. The diagnosis should never be difficult if one observes the rule to examine every infant after birth for a possible defect; but this condition, as well as others, is not infrequently overlooked through the inattention of the physician. We know of a case in which the attending physician ordered an enema for constipation in a newly born infant; the nurse administered it, according to her story, without effect; the child died a few days later, and at the post-mortem examination an imperforate rectum was discovered. In another instance the im-

perforate anus was not discovered till the sixth day, when the child's condition was too desperate to admit of surgical interference.

An imperforate œsophagus is necessarily fatal. Even a gastrotomy is out of the question on account of the inaccessible position of the infantile stomach. The diagnosis can be made with the aid of a soft rubber catheter. This should pass to a depth of 12 to 15 cm., in an infant of normal size, to enter the stomach. A failure to reach this depth indicates an obstruction in the œsophagus. (Tarnier.)

FIG. 5.



Anomalies of excess and deficiency in the number of the digits.
(IS. GEOFFROY SAINT-HILAIRE.)

ANOMALIES OF NUMBER AND EXISTENCE.—Abnormalities in the number of the digits is perhaps the most common example in this class.

The digits may be normal in number, but each lack a part. For instance, we have seen an infant lacking the terminal phalanges of all the toes. In another case we saw a remarkable

instance of heredity: the infant had supernumerary little fingers; its mother had had the same, and still showed the scars where

Fig. 6.

Supernumerary breast. Child nursed up to the twenty-third month from the gland upon the thigh, while another infant was nourished from the pectoral mammæ. (*Histoire des Accouchements*, p. 288.)

they had been removed. She asserted that several brothers and sisters and the paternal parent for two generations back had also exhibited this anomaly.

*Supernumerary Breasts* come under this head. These are more frequent than is generally supposed. Bruce, in an investigation of 3956 persons, found in 61 a supernumerary mamma or nipples—a proportion of 1.54 per cent. Leichtenstern places the frequency at 1 in 500. Both observers declare that men present the anomaly about twice as frequently as women. It is impossible to account for the accessory glands on the theory of revérsion, as they occur with no regularity in situation, but may develop at odd places on the body. The most frequent position is on the pectoral surface below the true mamma and somewhat nearer the middle line; but an accessory gland has been observed on the left shoulder over the prominence of the deltoid; on the abdominal surface below the costal cartilages; above the umbilicus; in the axilla; in the groin; on the dorsal surface; in the labium majus, and on the outer aspect of the left thigh.[1] In cases reported by Edwards and Handyside, heredity seems to have been a probable explanation for the development of supernumerary mammæ; but in the vast majority of cases no hereditary influence can be traced.

*Tail-bearing or Caudate Individuals* would come in this class. Ancient writers described human beings with tails like animals, but these accounts were long regarded as fabulous. From the middle third of this century, however, more than thirty cases have been described. It is said that they are quite common in certain parts of the world, as in Greece, Borneo, and, according to Freund,[2] in Alsace-Lorraine.

[1] W. A. Edwards: "Supernumerary Mammary Glands and Nipples," Medical News, March 6, 1886. A very good bibliography. See also Ahlfeld, op. cit.

[2] Virchow's Archiv, Bd. civ., p. 531.

From a study of the reported cases Hennig and Rauber[1] make the following classification:

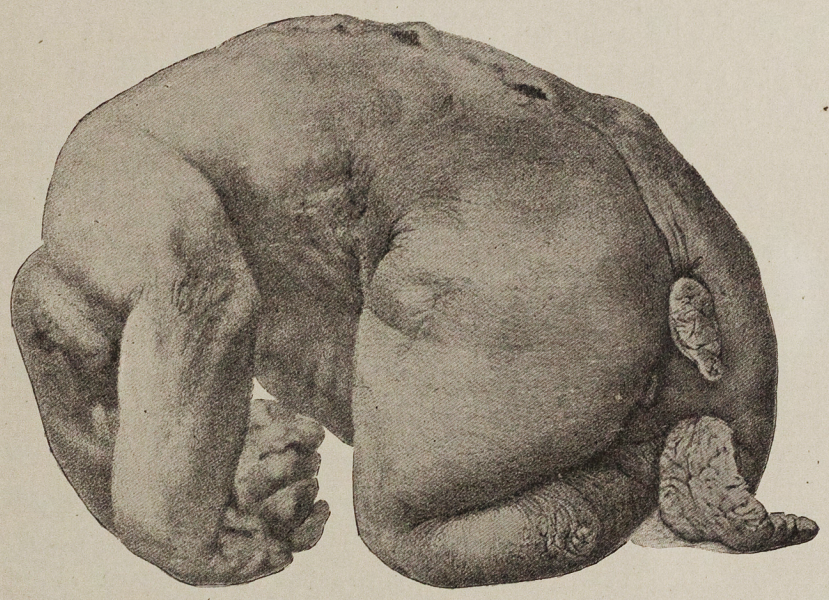
I. True Tails—Lengthening of the Spinal Column.

1. Containing vertebræ.

*a.* The usual number of coccygeal vertebræ.

*b.* More than five coccygeal vertebræ.

Fig. 7.



Infant with true caudal appendage. (Hennig and Rauber.)

2. Containing cartilage without definite divisions; (one case could, however, have been simply firm connective tissue).

3. Tail-like formation internally. (Freund's case, *loc. cit.*, is a good example.)

[1] Virchow's Archiv, Bd. cv., p. 83. A full bibliography.

II. Cutaneous and Lipomatous Tails.

1. Adherent. (Under surface fastened to body.)
2. Free.
3. A class in which the condition is somewhat indefinite and in which it may be difficult to say whether the growth is a tail-formation or some form of tumor.

Directly above the tail there is apt to be a depression, often surrounded by a growth of hair. The point of the tail is also usually provided with a hairy growth. In some cases the tail responds to irritation. In carefully dissected specimens there have been found a strong muscle (levator caudæ), superficially and above; other small muscles, more deeply placed and running in the long axis of the tail; muscle bundles running from the levator ani, and a caudal ligament.

## HETEROTAXIS.

By this term is meant the transposition of internal organs which is occasionally seen in the human species—*situs inversus viscerum*. The transposition may affect the organs of the thorax, but is more commonly seen in the abdomen. Rarely but a single organ is transposed; as Perls remarks, it is as though the internal organs were seen in a mirror. The heart may be on the right side, the apex directed toward the right; the cæcum upon the left side, the sigmoid flexure upon the right, etc.

This anomaly is said to be twice as frequent in men as in women. The individuals affected are usually right-handed. There is no interference with nutrition or function by the abnormal

arrangement of the parts. The diagnosis of the condition may be made during life, especially in the case of a transposed heart.[1]

## HERMAPHRODITISM.

True Hermaphroditism.—If one accepts Ahlfeld's definition of a true hermaphrodite—an individual with functionally active glands of both sexes, provided with excretory ducts—then a true hermaphrodite has not yet been discovered, and probably never will be. If, on the other hand, one allows an individual to be classed as a true hermaphrodite in whom there are sexual glands which histologically have the structure respectively of an ovary and a testicle, then there is a possibility of true hermaphroditism, for there has been at least one case carefully observed which would answer this description.

Klebs makes three divisions of true hermaphroditism.

I. Bilateral: on both sides, an ovary and a testicle.
II. Unilateral: on one side an ovary and a testicle, on the other an ovary or a testicle.
III. Lateral: on one side an ovary, on the other a testicle.

This classification, it is needless to say, is as yet theoretical. There are no undoubted cases of the kinds described to justify it. In very many instances true hermaphroditism has been claimed for an individual or a specimen, but very few indeed of these descriptions will bear scientific criticism.

Ahlfeld has collected quite a number of these cases from literature. Omitting all which are simply declared by the observer to be hermaphroditic without a histological examination of the glands, there remain:

[1] London Pathological Transactions, vol. xxviii. p. 448; Perls, Allgem. Path., Bd. ii. 323.

The case described by Barkow, in which there was undoubtedly one testicle, without, however, a vas deferens, and another body described as an ovary, which, histologically, was made up of fat, connective tissue, and bloodvessels.

The case of Berthold, in which there was a testicle in the right half of the scrotum; between the rectum and bladder a uterus unicornis; on the right side no adnexa, but to the left, round ligament, tube, and "ovary." The last lacked the characteristic histological elements of a normal ovary.

The case of Banon, in which to the left of the small uterus there was a tube and the ovary; to the right a testicle with vas deferens. The ovary again in this case showed no Graafian follicles; it was made up principally of connective tissue.

The case of Cramer-Meyer-Klebs, in which there was a testicle in the left scrotal sac; a uterus with tubes, parovaria, and round ligaments; to the right side an ovary, to the left a testicle. The description of this case is faulty. The diagnosis was declared by one of the writers to rest upon careful macroscopic and microscopic examination. Another says nothing about the latter; and Förster, in referring to the case, said that no Graafian follicles nor ovules could be discovered.

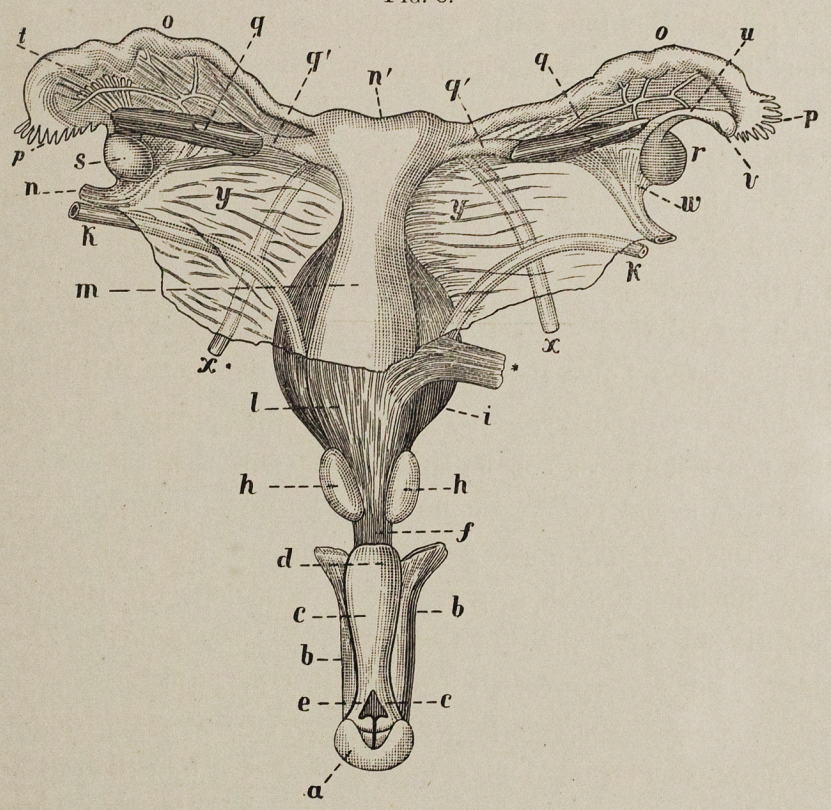
The cases of Gruber and of Klotz are too obscure to be of value. In one a cancerous mass was declared to be an ovary; in the other a cyst was taken for an ovary. In both cases there was a testicle upon the opposite side.

The case described by Heppner was that of a two-months'-old child. The external genitals were of the masculine type. The penis was imperforate. Internally, there was an infantile uterus with tubes and ovaries to either side. On each side also a testicle, separated from the ovary by the parovarium.

While the microscopic examination demonstrated the nature of the ovaries, it could not be demonstrated clearly under the

microscope that the neighboring glands were testicles. In addition to these cases collected by Ahlfeld, there has been quite recently described an interesting and important case of hermaphroditismus verus lateralis.[1]

FIG. 8.




Hermaphroditismus bilateralis. (HEPPNER.)

*a*, glans penis. *b*, corp. cavernosa penis. *c*, corp. cavernosa of urogenital canal. *d*, its bulb. *e*, its anterior arm. *f*, membranous part of urogenital canal. *h*, prostate. *i*, bladder. *k*, ureters. *l*, vagina. *m*, uterus. *n'*, fundus uteri. *o*, *o*, tubes. *p*, *p*, their infundibula. *q*, *q*, ovaries. *q'*, *q'*, ligaments of ovary. *r*, right testicle. *s*, left testicle. *t*, left parovarium. *u*, right parovarium. *v*, hydatid of Morgagni. *n*, *w*, bloodvessels. *x*, *x*, round ligaments. *y*, *y*, broad ligaments. *, muscle fibres from bladder and vagina.

[1] "Ein Fall von Hermaphroditismus," G. Schmorl, Virchow's Archiv, Bd. cxiii., p. 229.

Friedrich W., aged twenty-two years, art student from Berlin, sought admission to the surgical clinic in Leipzig, for a congenital defect of the sexual organs, which proved to be hypospadias. He desired an operation in order that he might gain the power of procreation and "be able to urinate from the end of his penis like other men." A closer examination showed that the scrotum was rudimentary in development. It began immediately in front of the anus and was divided in two by a raphé. On the right there was a small testicle; on the left, none. The penis was small, and drawn bow-like downward and inward. The glans was uncovered, well formed, but imperforate. On the under surface of the penis was a groove running backward 3.2 cm. to a small slit about ½ cm. long. Directly back of this the raphé of the scrotum began. The operation to correct the hypospadias was performed in two parts: first, to free the penis from its constrained position; second, to close the groove in it and make a urethra. An attempt to pass a catheter after the first operation failed to evacuate the bladder. On injecting fluid, a swelling was noticed in the left inguinal region. Finally the urine was drawn off by a Mercier catheter. After the second operation the same difficulty in catheterization was experienced, and again it was noticed that the left inguinal region became enlarged when fluid was injected into the catheter. Soon serious systemic symptoms developed, with swelling, redness, and pain in the left groin. The skin was incised in this region, and a body 5 cm. long and 2 cm. thick, with a band like the vas deferens, running into the abdomen, was exposed and removed. It was thought to be the left testicle. Shortly afterward the patient died. The post-mortem examination resulted as follows: Face bearded, hairs about 2 cm. long. Breasts undeveloped. Mons veneris had the hairy growth like a female, ending abruptly above. The penis, freed from its adhesions by

the first operation, measured 5.5 cm. in length on the upper surface, and had a circumference of 8 cm. The glans was 1.25 cm. long. To the sides of the penis were genital folds projecting above and grasping the penis between them. Internally there was discovered an opening into the urethra 3½ cm. back of the external orifice, where the colliculus seminalis usually lies, into which a sound could be passed for 15 cm. Further dissection discovered this canal to be a vagina and uterus, the latter separated into cervical and corporeal portions. On the left side the tube ran into the inguinal canal and was continuous with the body removed at the operation, which was found to be mainly the distended and distorted fimbriated extremity. Microscopic examination of this body, however, showed in it the remains of a sexual gland having all the histological characteristics of a fœtal ovary without ovules. On the right side were a round ligament, tube and ligament analogous to that of the ovary, all running down to the sexual gland in the right scrotal sac, which the microscope showed to be a testicle. There were no spermatozoa nor was there a vas deferens. It really seems that this might be called an example of true lateral hermaphroditism.

There remains still another case to be referred to in this category. Schmorl (*loc. cit.*) mentions an example of true unilateral hermaphroditism described by Gast.[1] It was a still-born baby with exstrophy of the bladder. There was a rudimentary but well-developed penis, which was perforated by a urethra and lay between folds of skin. The internal genitalia were more of the female type. The uterus was bifid; on the right it was solid and had a sheath-like extension composed of fibrous tissue. From the body of this uterine division there ran a tube 3 cm. long with an abdominal ostium. The left half

[1] T. Gast: "Beitrag zur Lehre von der Bauchblasengenitalspalte und vom Hermaphroditismus verus." Inaug.-dissert., Greifswald, 1884.

of the uterus, also solid, was hourglass-shaped, $2\frac{1}{2}$ cm. long and $\frac{3}{4}$ cm. broad. From this side also ran a tube with fimbriated extremity. To this was adherent an ovary showing follicles and ovules with nucleus and nucleolus. On the same side there was a testicle, the size of a pea, from which a ligament, the gubernaculum Hunteri, ran down to the base of the left scrotal sac. A microscopic examination showed this to be a testicle. On the right side no sexual gland could be found at all. This case can be called, therefore, one of true unilateral hermaphroditism—that is, if the account of it can be trusted. In all these cases there were anomalies in the external genitalia which will be described under the head of pseudo-hermaphroditism.

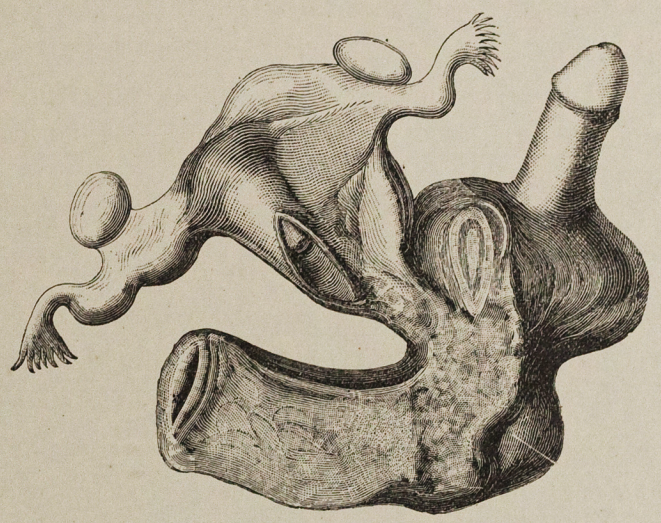
Pseudo-hermaphroditism.—As may be seen in the classification, individuals of this class have the glands of one sex, but other sexual parts either intermediate or mixed. They are in the vast majority of cases of the masculine sex, although this may be difficult to determine during life. Numerous instances are recorded of mistakes as to sex which continued throughout a great part or the whole of life. We have seen an individual don his first trousers at the age of nineteen, having been theretofore clad as a girl, until the beard began to grow and sexual inclination toward females began to be manifested.

A masculine pseudo-hermaphrodite has actually married as a woman, and only learned his true sex on consulting a physician for sterility. It is always safe in case of doubt as to the sex of an individual, to regard it as masculine.

There are many degrees of masculine pseudo-hermaphroditism, from a simple enlargement of the vesicula prostatica, without abnormality of the external genitals, to the full development of a uterus masculinus, divided into corporeal and cervical portions, with perfect tubes and a vagina opening externally into a urogenital cleft. In the latter case the penis is rudimentary

and there is hypospadias; the urethra opens by a separate canal at the urogenital cleft; there is a rudimentary development of the scrotal halves, and the testicles may be in the abdominal cavity. The vasa deferentia empty usually in the urethra, sometimes in the urogenital cleft, and rarely in the cavity of the vesicula. It is not strange that such creatures should be regarded as females,

FIG. 9.



Spurious hermaphroditism. The round bodies are testicles.

for the only distinctive mark of their true sex may not be discoverable till after death. In intermediate grades the external genitals may not be much affected, and the uterus masculinus and vagina may open into the urethral canal of a fairly well formed penis. The scrotum may show various grades of development: one-half may be pretty well formed and contain a testicle, while the other is rudimentary and empty. The testicle in such a case may be detected in the inguinal canal or may be altogether in the abdomen. However doubtful might be the sex of a masculine pseudo-hermaphrodite during life, an examination of the pelvic cavity after death should settle the matter definitely.

And yet mistakes have been made in the description of post-mortem specimens. There was reported to the New York Obstetrical Society, March 1, 1887, a case of so-called true hermaphroditism, which was accepted unchallenged, and yet there was no microscopic examination of the sexual glands, and the bodies which were designated as ovaries are evidently nothing but convolutions of the tubes attached to the uterus masculinus. This was undoubtedly a case of spurious hermaphroditism in the male.

Feminine pseudo-hermaphroditism is rare, and reported cases should always be regarded with the suspicion that the individual is really a male. A hypertrophied clitoris, perhaps a rudimentary vagina, ovaries prolapsed into the labia, the formation of a scrotum and the existence of vasa deferentia are the characteristics of this class.

The following remarkable case was reported to the *New York Medical Journal*, Nov. 22, 1890, by Dr. C. W. Fitch, at one time in charge of the sanitary service of Salvador, C. A.:

"J. H. A., a house-servant, of masculine features and movements; aged twenty-eight years; height, five feet seven inches; weight, one hundred and thirty-nine pounds; was arrested by the police for violating the law governing prostitution. On examination, both female and male organs of generation were found in a remarkably well-developed condition. The labia majora were of normal size, but flattened on their anterior surface. The labia minora and hymen were absent. The vagina was spacious, four inches and a quarter long anteriorly and six inches posteriorly. The os uteri was torn on the left side. There was profuse leucorrhœa. Seven years before she had given birth to a normal female infant. In place of the clitoris there was a penis, which, when in erection, measured five inches and a quarter long by three inches and five-eighths in circumference. The glans penis

and the urethra (?) were perfectly formed. The scrotum, which was two inches and an eighth long, contained two testicles (?) about an inch in length and two inches and a half in circumference. The mons veneris was sparsely covered with short, straight, black hair. Both sets of organs were perfect in their functions, semen being ejected from the penis (?) and the ovaries being capable of producing eggs. Scanty menstruation occurred every three weeks, and lasted but two days. Sexual gratification was said to be equally distributed between the two sets of organs." Stripped of inaccuracies, this is a description, doubtless, of a remarkable example of pseudo-hermaphroditismus femininus.

## MONSTROSITIES.[1]

### SINGLE MONSTERS.

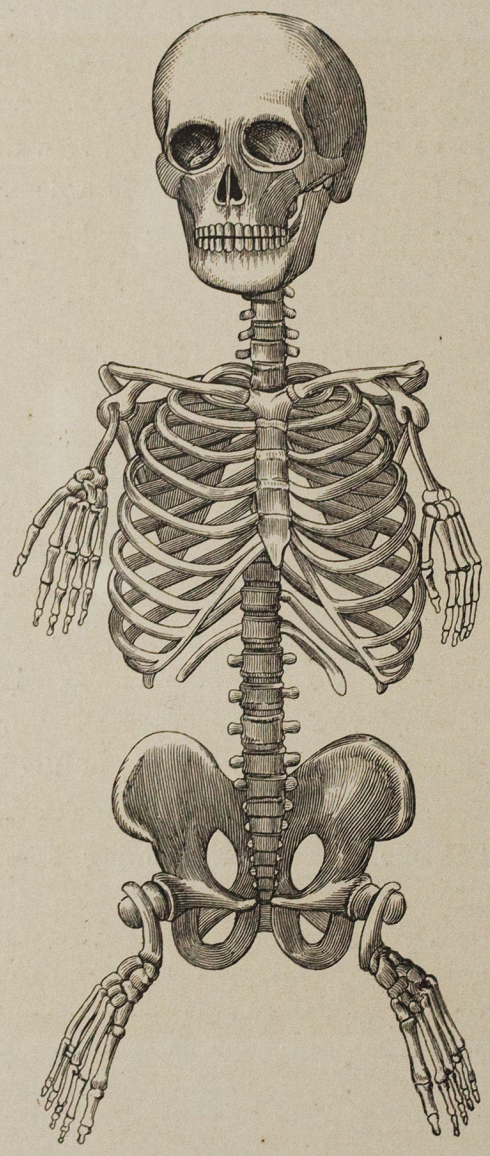
ORDER I. AUTOSITIC SINGLE MONSTERS, capable of an independent existence. The characteristics of this order are mainly arrest of development, fusion, and displacement of important parts of the body.

ECTROMELUS.—The word signifies an aborted or imperfectly formed limb. Examples of this anomaly show every degree of arrested development, from entire absence of the limb to simple shortening. Three divisions of this species are made by Geoffroy Saint-Hilaire, to which, following Förster, we add a fourth (micromelus).

[1] The word is used in its scientific sense to denote certain exaggerated deviations from normal development, which give to the individual a strange or even inhuman appearance. In this sense the word is firmly imbedded in modern medical nomenclature. Like some other medical terms of ancient origin, the first significance is entirely lost, while the word itself is retained. A good example of this is the membrana decidua serotina of Hunter.

1. *Phocomelus.*—Derived from φώκη, seal and μέλος, limb. The distinctive mark of this class is an exaggerated shortening and rudimentary development of the long bones of the limbs, while the feet and hands may be normal in development, looking as

FIG. 10.

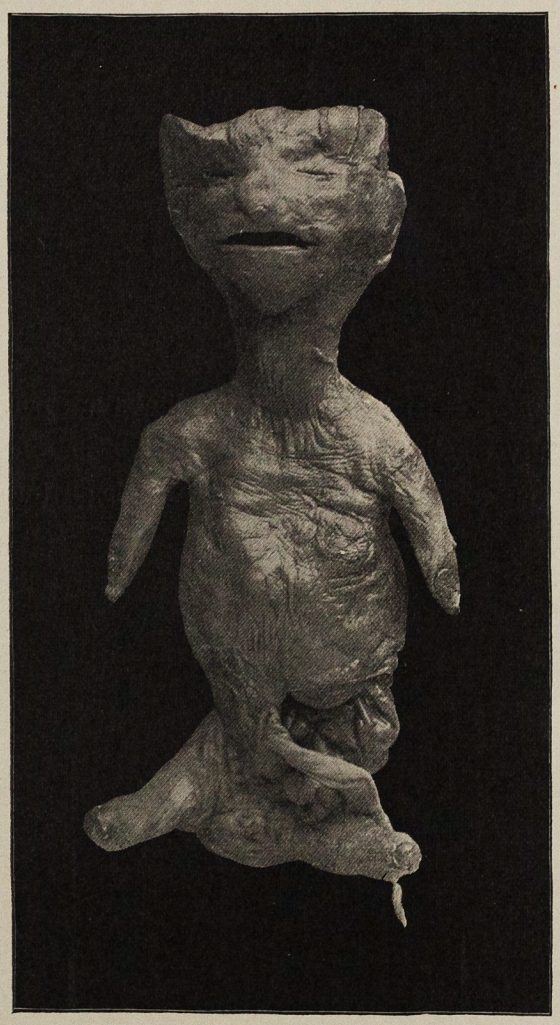


Skeleton of a phocomelus. (Musée Dupuytren.)

though they sprang directly from the shoulders and the hips. (See Plate V.)

2. *Hemimelus.*—Under this name are described cases in which the lower portion of the limbs is very ill-developed or altogether absent.

FIG. 11.



Hemimelus.

The specimens of this class, however, are apt to be not so distinctive as the phocomeli. (See Plate VI.) Perhaps but one

limb is affected, and there may be traces of the three divisions in a rudimentary form.

For these cases the term "peromelus" (perobrachius, peropus) is employed by Förster.

Fig. 12.

Peromelus.

3. *Micromelus.*—In this variety the limbs are normal in form but abnormally small. Rarely a single limb is affected or the abnormality is confined to the arms or legs (microbrachius, micropus).

Fig. 13.

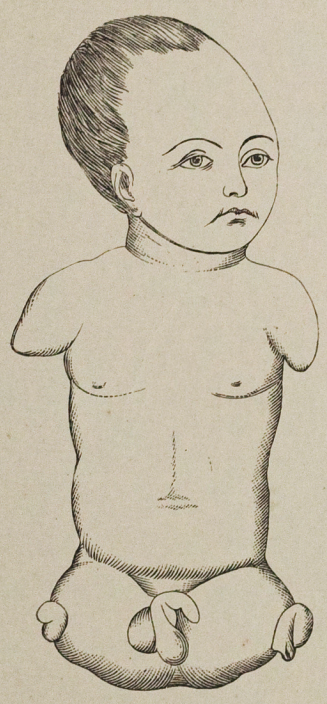
Amelus.

4. *Ectromelus.*—In a typical case of ectromelus there is an entire absence of limbs (amelus). Sometimes small stumps mark the places for arms and legs.

The abnormality may be confined to the upper extremities (abrachius) or to the lower extremities (apus, monopus).

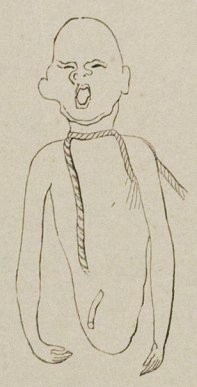
These cases are the result of arrested development, and are to be distinguished from intra-uterine amputations by amniotic bands.

Fig. 14.



Ectromelus.

Fig. 15.



Apus.

Fig. 16.

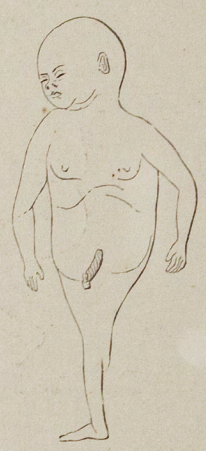
Monopus.

Symelus.—The symeli are characterized by imperfect development of the pelvis and lower extremities; by *atresia ani et urethræ;* by more or less intimate fusion of the lower extremities, and by a twisting of the lower limbs so that the femora are united by the external condyles, the legs by the fibulæ, and the feet, if they exist, by the fibular edge and little toes, so that the heels look forward. Geoffroy Saint-Hilaire makes three divisions of this species.

1. *Symelus.*—There is more or less fusion of the lower limbs, but two feet are to be distinguished.

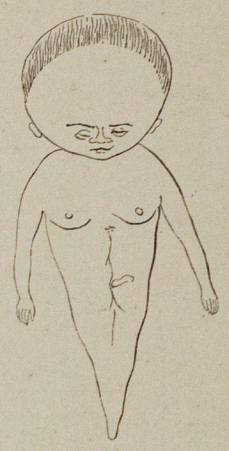
2. *Uromelus.*—The fusion of the limbs is more complete and there is but a single foot. The thigh usually shows evidence of double formation, but the leg is single. The foot may be normally formed, imperfect, or may possess six to eight toes. In the last case the big toe is always in the middle, and the heel in all forms is turned forward.

FIG. 17.



Uromelus (Sympus monopus, FÖRSTER).

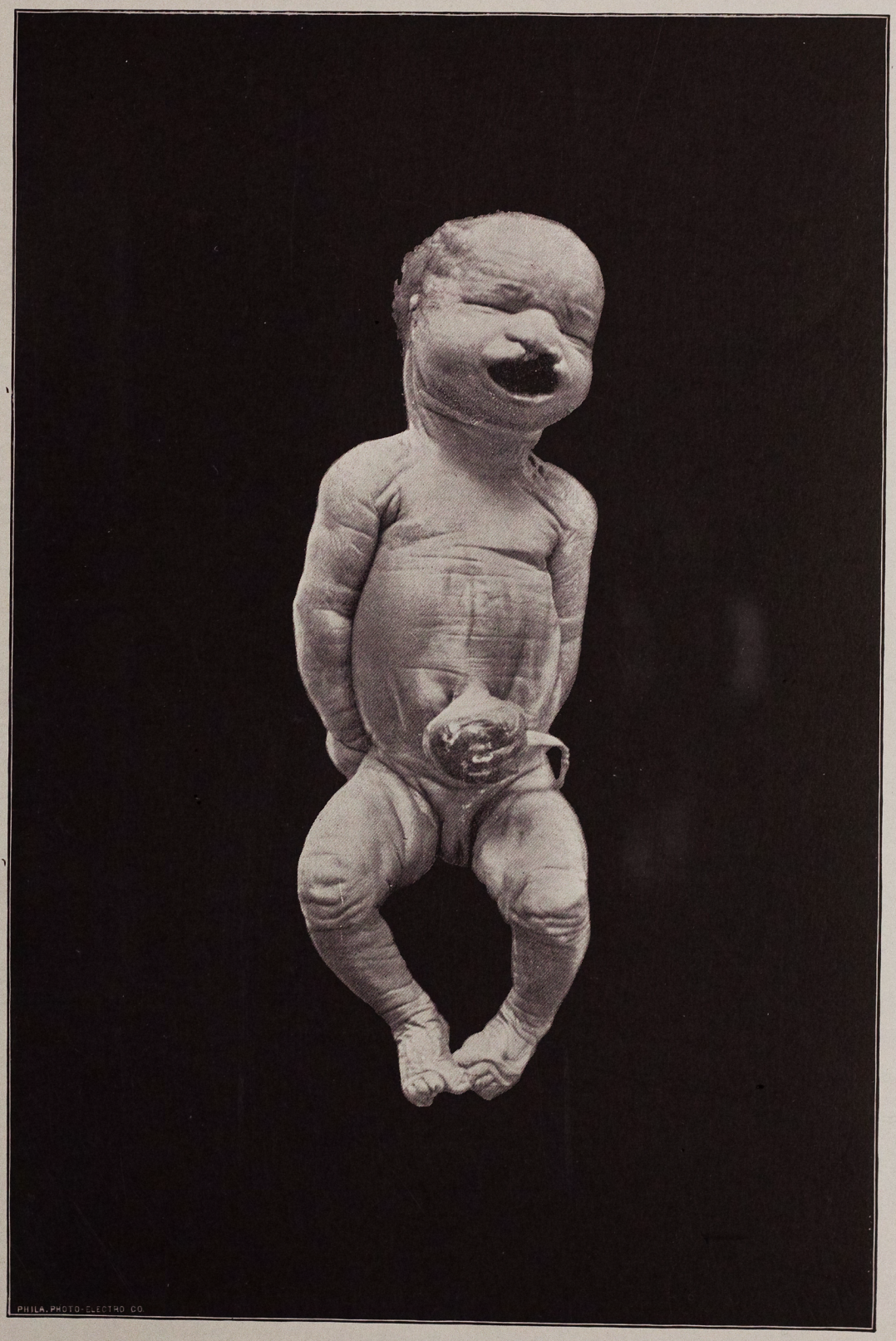
FIG. 18.



Sirenomelus (Sympus apus, FÖRSTER).

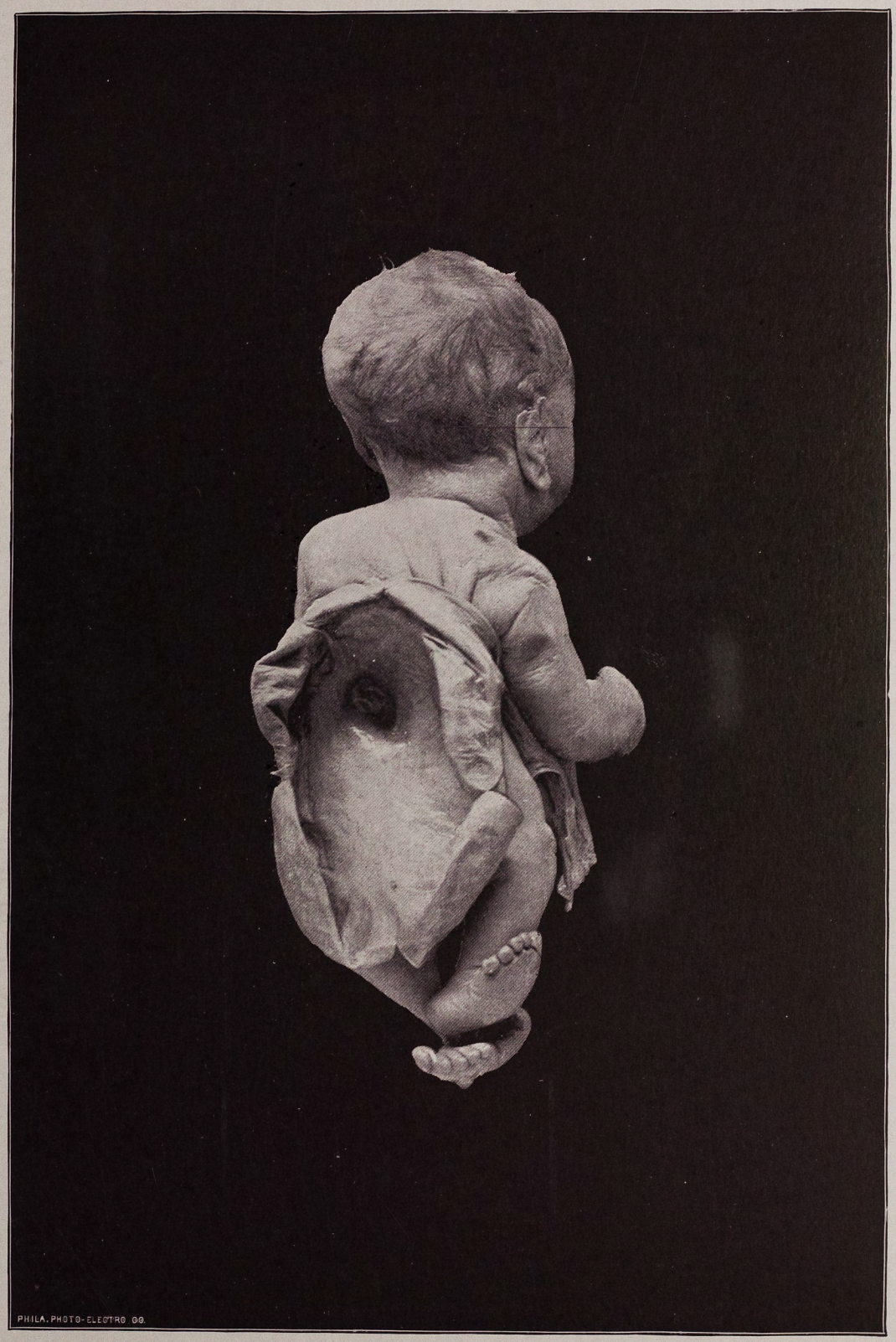
3. *Sirenomelus.*—In this form the lower extremities are intimately fused. There is but a single femur, showing, perhaps, double formation at the lower extremity. The leg has only one bone and there is no trace of a foot, or, at most, but a single toe. The pelvis is defective in development, and the bones, especially the sacrum, are twisted in a remarkable manner. (See Förster, p. 67.) The rectum and lower end of the colon, the bladder, and sexual organs, are usually absent. (See Plate VII.)

PLATE I.




UMBILICAL HERNIA OF MODERATE DEGREE
AND DOUBLE HARE-LIP.

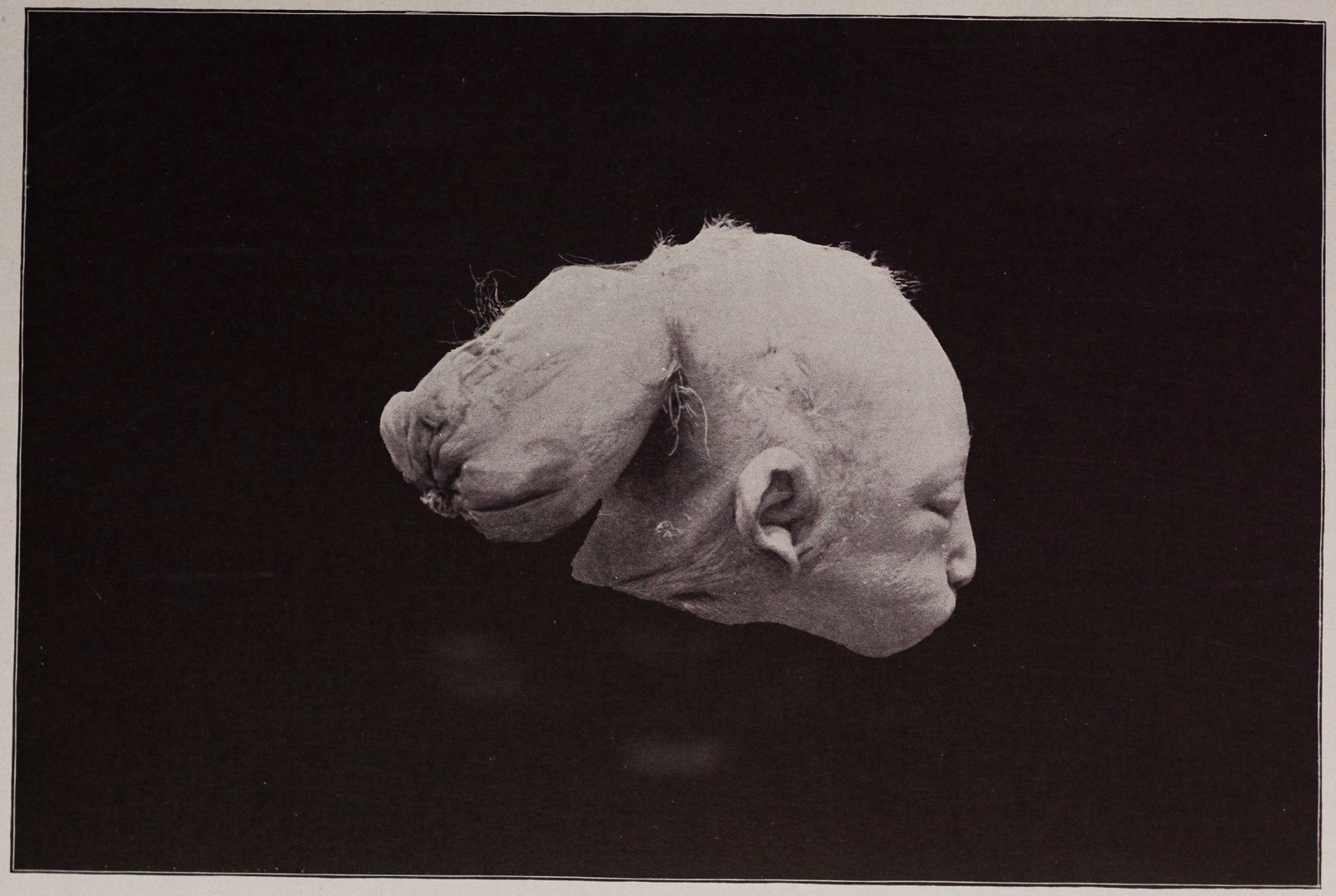
PLATE II.




SPINA BIFIDA

WITH LARGE SPINAL MENINGOCELE.

PLATE III.



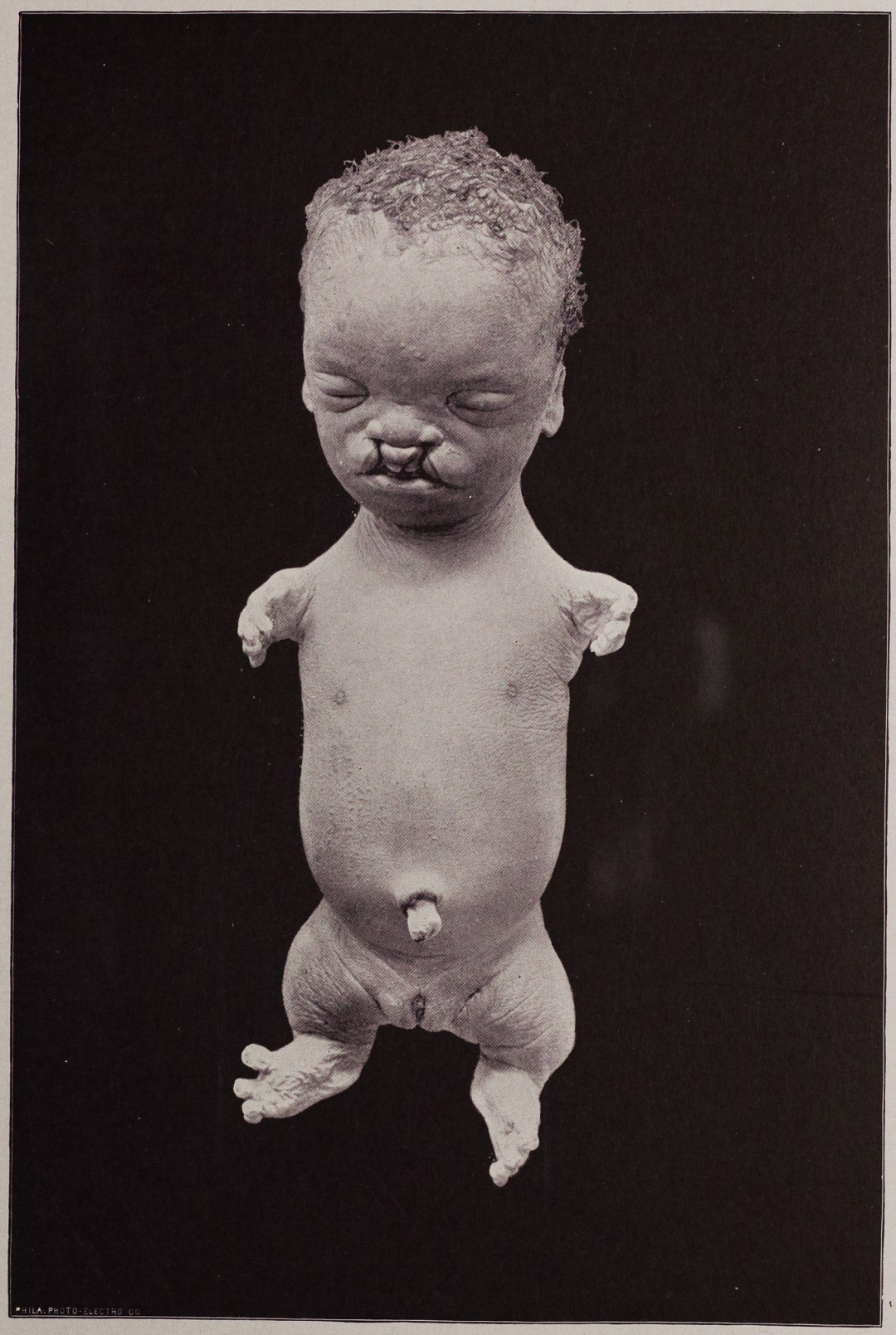
LARGE ENCEPHALOCELE.

PLATE IV.

DOUBLE ENCEPHALOCELE.

(CEPHALIC MENINGOCELE.)

Plate V.

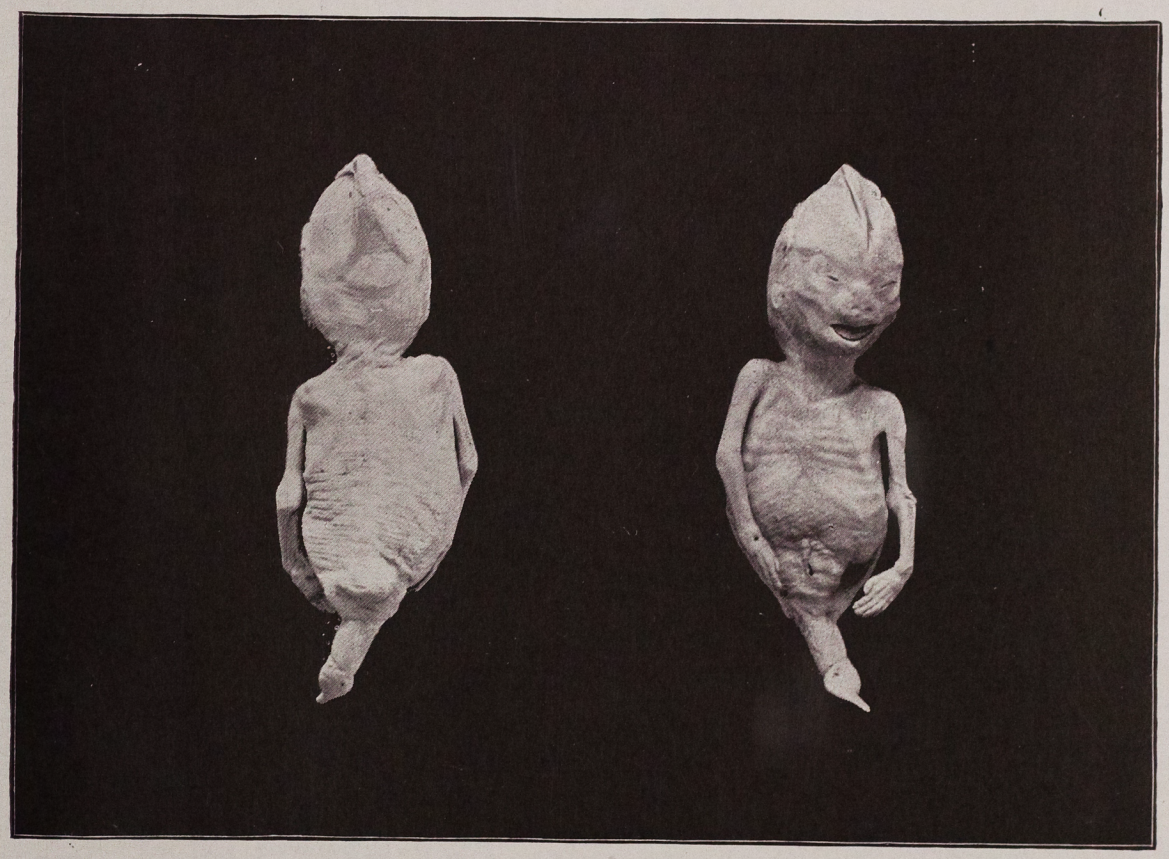



PHOCOMELUS.

PLATE VI.


HEMIMELUS.

PLATE VII.



SIRENOMELUS.

(FRONT AND REAR VIEW.)